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Cerebral Palsy Bulletin

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BIOGRAPHY

PROF. ALBRECHT PEIPER. *With frontispiece.* By Heinz Prechtl 1

EDITORIALS

HEARING AND SPEECH DISORDERS. O. L. Zangwill 3
DELAYED SENSORY FEED-BACK IN THE STUDY OF DEFECTIVE SPEECH AND WRITING. H. Kalmus 5
PROGNOSTIC VALUE OF NEUROLOGICAL EXAMINATIONS IN THE NEWBORN. R. W. Smithells .. 7
GLUTAMIC ACID FOR MENTAL DEFECT? D. Russell Davis 10
PHYSICAL TREATMENT OF CHILDREN WITH CEREBRAL PALSY J. H. Crosland 11
CEREBRAL PALSY IN SOUTH AFRICA. Ross Mitchell 14
LETTERS AND QUESTIONS PLEASE. The Editors 16

ORIGINAL ARTICLES

The Subacute Encephalitides. Prof. Ludo van Bogaert, M.D. 17
Piaget's Approach to Problems of Intellectual Development. Anne-Marie and Joseph Sandler .. 25
Problems of Hypoxia in the Genesis of Cerebral Palsy. J. B. Brierley 29
Lower Extremity Bracing in Cerebral Palsy. Russell V. Fuldner 34
The Child's Hearing for Spoken Language. Mary D. Sheridan 39
Hearing and Speech Disorders in Childhood. Gavin Livingstone 46
Hearing and Speech Disorders. Michael Reed 52
A Description and Classification of Common Speech Disorders associated with Cerebral Palsy. T. T. S. Ingram and Jane Barn 57

SPECIAL ARTICLES

L'Hôpital des Enfants Malades, Paris. Léon Tixier (with cover picture) 70
What's to be Done? A Real-life Problem and Some Replies.
The Problem: Guy Tardieu 71
and Some Replies: W. J. W. Sherrard 72
Gavin C. Gordon 72

LETTERS TO THE EDITOR

Is there a Syndrome of Brain-damage in Children? Kenneth Cameron 74
Melvin Lewis 75
Frederick H. Stone 76
Autofaradisation for Paretic Muscles. F. Vèle 77
Growth Control of the Sympathetic System by a Specific Protein Factor. Rita Levi-Montalcini .. 78

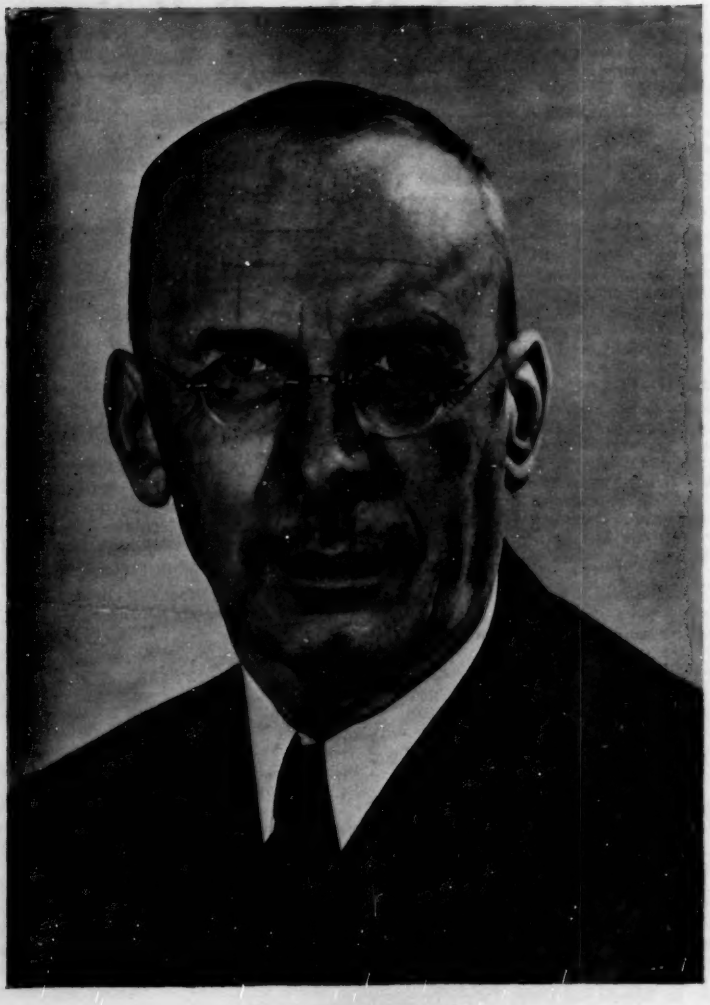
BOOK REVIEWS

Physical Disability—a Psychological Approach. By Beatrice A. Wright (Mrs. E. M. Mason) .. 79
Health in Childhood. By Prof. R. W. B. Ellis. (B. S. B. Wood) 80
Mirage of Health. By René Dubos. (Roy M. Acheson) 81
Psychological Problems in Mental Health. By S. B. Sarason and Thomas Gladwin. (T. T. S. Ingram) 82
Understanding and Teaching the Dependent Retarded Child. By Louis E. Rosenzweig and Julia Long. (Grace E. Woods) 83
Baby Talk. By Morris Val Jones. (Mary D. Sheridan) 84
An Introduction to Embryology. By B. I. Balinsky. (Roger Warwick) 84
Pathology of Infancy and Childhood. By Agnes R. Macgregor. (N. R. Rowell) 85
Congenital Deformities. By Gavin C. Gordon. (R. B. Zachary) 85
International Catalogue of World Mental Health Films. Ed. T. L. Pilkington. (Jonathan Gould) .. 86

NOTICES

Gadgets for Handicapped Housewives. M. R. Morgan 6
Back Numbers, Bound Volumes and the 'Clinics'. E. Clayton-Jones 13

ABSTRACTS 87



Prof. Albrecht Peiper

PERSONALITIES OF TODAY

Prof. Dr. Albrecht Peiper

By HEINZ F. R. PRECHTL, M.D.

PROFESSOR PEIPER is a children's specialist. His father was a professor of paediatrics, and so he grew up in the atmosphere of child medicine. He studied under Czerny

The professor studying the mimeographed discussions in the lecture-room at Gröningen with his charming wife.

at the Charité in Berlin. As a young assistant he already began to take an interest in the physiology of the mind and the functions of the nervous system in newborn babies and young children. With an admirable constancy of purpose he has devoted his whole life to this field of activity, and it is in this sphere that he has done his most impressive life-work.

When Peiper started his scientific work, there were philosophical and psychological speculations on brain activity in children, but very few scientific analyses had been made. Preyer had published his famous diary on the development of his child, but right from the start, as his teacher Czerny had said of him, Peiper had always wanted to 'prove everything for himself and not rely on the findings of others'. It is to this quality that we owe his 150 publications on research into the brain function of children.

The background on which Peiper based his own research on children is the classical neurophysiology of Sherrington, Magnus, Rademaker and Pavlov. Later on came the works of von Holst on central automatisms and ethology as developed by Lorenz and Tinbergen.

There can hardly be anyone else with a

The Moro response being demonstrated at the Gröningen meeting in 1960.

similar knowledge of the literature concerning the brain function of children from antiquity to the present day. It is this encyclopaedic knowledge and his own work that has given us the solid basis on which we can now build diagnostic neurological methods of research into the cerebrally damaged small child.

For all those working on the activity of the nervous system in children, whether they be paediatricians, neurologists or child psychologists, his book, *The Peculiarity of Brain Function in Children*, is a standard encyclopaedic work, not merely a summary of the opinions of one particular school.

Hearing and Speech Disorders

HUGHLINGS JACKSON is said to have remarked that neurologists, like SHAKESPEARE'S Dogberry, seem to believe that reading, writing and talking come by nature. Although the great man doubtless had his tongue in his cheek—if indeed he ever said any such thing—it is certainly true that neurologists have tended to stress constitution rather than environment, nature rather than nurture. This outlook has almost certainly been responsible for the widespread lack of interest among neurologists in the problem of learning and may well have engendered a certain fatalism towards educational handicap. Fortunately, there are signs of a change in attitude: failures in learning of the kind which produce backwardness in language or manipulative skill are attracting increasing attention and stimulating fresh remedial efforts. This is only a beginning, it is true, but it is none the less an important beginning.

With due respect to Dogberry, it is surprising how little is really known about the acquisition of speech. It is plain, however, that any condition limiting auditory perception in infancy is liable to retard or distort its development. For this reason, the need for early detection of hearing loss takes on particular importance. In his admirable report on hearing and speech disorders, Mr. GAVIN LIVINGSTONE makes clear that an appreciable proportion of babies must be considered 'at risk' from the standpoint of hearing loss (and consequently of impaired speech development) and rightly stresses the need for an appropriate register to be kept available and as complete as possible. At the same time, neither he nor Mr. MICHAEL REED (p. 52), who approaches the problem from the standpoint of the clinical psychologist, has any illusions about the difficulty in ascertaining hearing loss in children too young for audiometry. Both are agreed on the need for repeated observation and study under a variety of conditions; both, too, stress the need for team-work. One would not wish in any sense to disagree, but how helpful it would be if these and other experts would formally define the criteria on which their assessments of hearing loss are based! Their experience might then be put to good advantage in the development of more objective methods of examination.

Since EWING'S work some thirty years ago, it has been known that many children presenting the picture of 'congenital auditory imperception' are in fact afflicted by high-frequency deafness. As Dr. MARY SHERIDAN points out in her lucid account of hearing in relation to speech (p. 39), it is the consonant sounds that are mainly affected; consequently a young child may begin to vocalise and use vowel sounds at a normal age, and for this reason the hearing disability may for long remain unrecognised. One might suggest that phonetic analysis of the child's earliest speech sounds might well prove valuable in the early diagnosis of hearing loss.

In spite of the obvious importance of auditory handicaps for speech retardation, there are clearly many forms of developmental speech disorder that are independent of sensory loss. Indeed, as

Mr. REED points out, it is the children with communication disorders *not* due to deafness who receive the least attention from our educational system. These disorders have been well described by Dr. T. T. S. INGRAM in an earlier article (this *Bulletin*, Vol. 2, 1960, p. 255), which should do much to rearouse interest in their classification and study. Of more immediate concern, however, are the varieties of speech defect presenting in cerebral palsy, which form the subject of yet another comprehensive review by Dr. INGRAM, this time in collaboration with Miss BARN (p. 57). These authors point out that whereas dysarthria is of course common in diplegic or dyskinetic states, retardation in speech secondary to mental impairment is of far greater importance. Indeed it is suggested that language disorder in cerebral palsy generally is due not to defect of a specific kind (aphasia) but to retardation consequent on slow learning and poor intelligence. If this proves to be the case, it would indicate that there is an important difference between developmental disorders of language and aphasia due to left-sided brain injury sustained after the acquisition of speech.

Dr. SHERIDAN's plea that the whole question of defective language in cerebral palsy needs review should be widely endorsed. As she rightly points out, much time and energy have been devoted to remedial procedures obscure in rationale and dubious in outcome. Although speech teachers undertake much important and devoted work, they at present receive little guidance from doctors or psychologists and are given little opportunity to place their experience and methods at the disposal of research workers. One may therefore hope that solid support will be given to Dr. SHERIDAN's proposal for a concerted scheme of research on speech problems under the leadership of an experimentally-minded paediatric neurologist. Such a research scheme would draw on experience in the paediatric, psychological and educational fields but should be firmly built on neurological foundations.

O. L. ZANGWILL

Delayed Sensory Feed-back in the Study of Defective Speech and Writing

MANY otherwise normal children suffer from speech defects or writing difficulties, and such defects are of course part of the picture in many handicapped children. In most of such cases the faulty function is localised in the nervous system—either in the motor or the sensory field, or centrally. It is difficult to imitate such defects experimentally, for instance by drug action, and it might therefore be interesting to explore them by a slightly indirect approach.

One such approach, provided by electronic developments started during the war, consisted in delaying the acoustic feed-back to a person while he was talking. This gross disturbance of the monitoring of his speech produces stammering.[1] At first the method was used to unmask recruits who were pretending to be deaf. It proved impossible for a hearing person to escape the conflict between the proprioceptive information coming back from his larynx during speech, and the acoustic feed-back provided by replaying through ear-phones a recording of his own speech, with delays of 0.1–0.5 seconds. Deaf people, however, were wholly undisturbed by this arrangement. The apparatus for producing acoustic delay was later used to produce rather striking effects on some non-vocal activities,[2] such as tapping, banging or the playing of musical instruments. Among other things it was found that the effect was sometimes to add one to an intended sequence of knocking signals—for instance 6 knocks were produced instead of 5.

Recently an apparatus was built which delayed the visual performance of a person's writing activities by similar amounts of time. Again the conflict between the undelayed kinesthetic information from the writer's hand and the delayed feed-back from the retina and probably the eye muscles resulted in gross disturbances of writing and some related activities, such as drawing and tracing.[3]

It seems that people subjected to acoustic or visual delay try to cope with their difficulties in two alternative and conflicting ways. Either they try to maintain their rate of speech or writing, aiming at completeness but sacrificing quality in detail; or, they aim at accuracy in detail, which forces them to slow down and to leave out certain features altogether or slur them quite badly. The degree to which these tendencies are 'traded' against each other varies from person to person and also according to instructions and other experimental conditions. It is not suggested that any of the pathological features of speech defect or writing difficulty are directly comparable to these effects of delayed sensory feed-back in normal adults; but it would be

1 Lee, B. S. *J.acoust.Soc. Amer.* 1950, 22, 825.

2 Kalmus, H., Denes, P. and Fry, D. B. *Nature, Lond.* 1955, 175, 1078.

3 Kalmus, H., Fry, D. B. and Denes, P. *Language and Speech.* 1960, 3, 96–108.

interesting to investigate, by means of these delays, the stability or otherwise of the speech or writing of various defective patients. This might occasionally help in differential diagnosis and might even influence the choice of therapy. A necessary basic investigation is that of the effects of delayed feed-back on speech and writing as these faculties develop in normal children.

H. KALMUS

NOTICE

GADGETS FOR HANDICAPPED HOUSEWIVES

A USEFUL little booklet,* compiled in Sweden and now on sale there and in New York, was originally produced to help in the replanning and rehabilitation of the housewife who has become physically handicapped in some way and has had to adapt her domestic life to fit in with her reduced mobility or dexterity.

The booklet has in fact some very useful suggestions for any woman—or for that matter any man—who is handicapped but who aims at achieving a maximum of independence at home. Some of the gadgets described and illustrated are well known in this country, and the authors mention the excellent work being done in this field at King's College Hospital and the Middlesex Hospital in London. It is, however, helpful to have such clear photographs and illustrations gathered together in one book, and the section on gadgets and adaptations is conveniently divided into those designed for the chairbound and those for people with restricted hand and arm movements. There are also useful chapters on kitchen and bathroom layouts, designs for fittings, sanitary equipment and electrical installation, training in various facets of domestic work, advice on food hygiene and some general tips on exercises for the handicapped and often housebound woman.

The ideas and suggestions are sensible and practical, and the emphasis throughout is on simplicity and economy, the authors fully appreciating that much of the specialised equipment is expensive and often beyond the means of the normal family. A good illustration of the commonsense outlook is the photograph on p. 6, where a chairbound woman is preparing food at a bench with all the jars and bottles placed horizontally on the shelves with the caps towards her for easy handling—a very simple device, but one which would not immediately come to mind without such a clear illustration.

M. R. Morgan

* The Physically Handicapped Housewife. S.V.C.K.'s Publication Series, No. 6, 1959, pp. 68, $1. Obtainable from The National Society for Rehabilitation of the Disabled, 701 First Avenue, New York 17, N.Y., U.S.A.

Prognostic Value of Neurological Examinations in the Newborn

IT was an overcast July day last summer when Dr. JAN DIJKSTRA presented his thesis* and successfully defended it against the assaults of his medical and academic colleagues. Members of the Little Club study group who were in Gröningen at the time enjoyed his victory with him. His subject was a follow-up study of infants examined neurologically soon after birth by HEINZ PRECHTL head of the Section of Experimental Neurology in Gröningen.

The infants were born between 1954 and 1957 in the same obstetric unit, and were assessed as normal or abnormal in three distinct respects. On the basis of histories supplied by obstetricians, those with prenatal or paranatal complications (128) were distinguished from those without (25). The complications were: haemorrhage during pregnancy, mild to serious toxaemia, postmaturity (about 3 weeks over term), prolonged labour, slow (below 70 min.) or irregular foetal heart, forceps delivery, breech presentation, version and extraction, Caesarian section and postpartum asphyxia. Dr. PRECHTL classified the infants in the neonatal period as neurologically normal or abnormal, the abnormalities including lateralising signs, diminished or increased tone, diminished or increased activity, and absence of certain reflexes. All the infants with normal obstetrical histories were found to be neurologically normal. Nine infants with perinatal complications were untraced. Of the remaining 119, 60 had been deemed neurologically abnormal.

Finally, the infants were assessed at Dr. DIJKSTRA's follow-up examination. He saw 116 infants from the perinatal complications group (3 had died of neurological disorders) and 20 of the 25 controls at ages ranging from $1\frac{1}{2}$ to 4 years. He took a detailed history from the mother, made neurological, psychological and developmental assessments, and recorded an EEG.

The greater part of his thesis was concerned with an attempt to show in which respects obstetric complications, neonatal neurological findings and abnormalities found at follow-up may be correlated. The relationship between the first two groups seems clear. When there is perinatal hypoxia, about two-thirds of infants show neurological abnormalities in the neonatal period. Other obstetrical factors have no independent effect and are only related to neurological disturbances by virtue of their ability to cause hypoxia.

Of the 20 normal controls seen at follow-up, 3 had had postnatal illnesses potentially capable of causing brain-damage and were discarded. The remaining 17 were normal. It is unfortunate that the control group was so small but this probably does not affect the main conclusions. For similar reasons, 9 children were dropped from the group with perinatal complications; 2 had died with epilepsy and one with

* De Prognostische Betekenis van Neurologische Afwijkingen bij Pasgeboren Kinderen. University of Gröningen M.D. Thesis, 1960.

hydrocephalus, leaving 110 for follow-up study. Forty of these were considered to have neuropsychiatric disorders 1½ to 4 years later. Of these 40, 36 had been considered neurologically abnormal in the neonatal period. A further 21 who had been abnormal at birth were normal at follow-up.

Analysing in more detail the kinds of abnormality found in the neonatal and follow-up examinations, Dr. DIJKSTRA found a strong relationship between the neonatal hyperexcitability syndrome (*Overprikkelbaarheidssyndroom*) and the later appearance of the choreatiform syndrome. The former consists of hyperactivity, a readily elicited Moro response, and low-frequency, high-amplitude tremor. The choreatiform syndrome, which appeared later in 54 per cent of these children, is characterised by hyperactivity, short attention span, mood instability and learning difficulties, especially with reading and arithmetic.

To summarise, this part of the thesis showed that a history of perinatal hypoxia *or* the detection of neurological abnormalities in the neonatal period carries a two-to-one risk of subsequent abnormality, though this is more commonly a disturbance of behaviour than of neuromuscular function. When lateralising signs were detected in the neonatal period they were often still present at the follow-up examination. In contrast, when the nervous system appears normal at birth there is a very high probability that the child will remain normal.

Electroencephalography did not produce much helpful information. Observation of the children's behaviour generally confirmed the mother's account and emphasised the restlessness of many children who had been hyperactive in the neonatal period.

The results of developmental tests were confusing. Children under 2 years at follow-up were tested by RUTH GRIFFITHS' methods and no difference was found between those who had been neonatally normal and those who had been abnormal. Children over 2 years were given the '*Kleinkindertest*' of BÜHLER and HETZER, and here significant differences were found. However, if a D.Q. of 100 is taken as average normal, the differences shown arose because 72 per cent of the neonatally normal group achieved scores above this level.

Dr. DIJKSTRA tried to determine whether differences in behaviour and developmental level in the various groups might have been attributable to postnatal environmental factors. No differences could be found between the groups in respect of the parameters recorded.

The children with neonatal neurological abnormalities walked at the same age, on average, as the neurologically normal group, but they seemed slower in acquiring sphincter control. This is presumably another facet of the distractable behaviour of children with the choreatiform syndrome.

Comment

This thesis represents a serious attempt to answer some difficult questions. It is not easy to read, being somewhat overburdened with figures, tables and charts, and there are some numerical misprints in the original Dutch text. The *chi*-squared test has sometimes replaced reason—for example, where breech delivery is shown to be safer than vertex delivery if there is toxaemia or haemorrhage. It must be constantly borne in mind that the conclusions reached relate to the neonatal neurological examination of one particular

(highly experienced) observer. There is not yet wide enough agreement on techniques and interpretation to expect closely similar results from other centres.

Within these terms of reference there emerge some solid facts. Obstetrical complications endanger the infant's brain by causing 'hypoxia'. The more convincing the evidence of hypoxia, the greater the likelihood of brain-damage. If no neurological abnormality is found in the neonatal period, later examination is very unlikely to reveal any neurological or psychological disorders. If lateralising signs are found in the neonatal period they are very likely to persist. The most common pattern of disturbed neurological function in the neonatal period is the hyperexcitability syndrome. This correlates closely with hyperactive, distractable, mood-labile behaviour in later childhood.

Like most research projects, this study raises more questions than it answers. One would like to know whether some of these restless children, who were restless in infancy and restless in the neonatal period, ran short of oxygen because they were restless foetuses. But perhaps it is easier to oxygenate them than to tranquillize them before they are born?

R. W. SMITHELLS

Note: I am grateful to Dr. Jan Dijkstra for elucidating in correspondence some of the problems that arose in reviewing his work.—R.W.S.

Glutamic Acid for Mental Defect?

SEVERAL circumstances raised hopes in the 1940's that glutamic acid would improve the intellectual performance of severely subnormal patients. Følling's discovery in 1934 of phenylketonuria had given a fillip to the belief that the causes of severe subnormality would be found to lie in errors of metabolism. Interest in the metabolism of brain tissue *in vitro* had been at a peak in the 1930's, and glutamic acid has been found by Weil-Malherbe to be the only amino-acid capable of maintaining the oxygen uptake of sliced brain tissue. The remarkable successes of German troops in the campaigns of 1940 had revived irrational beliefs that the performance of men, as of cars, could be boosted by additives to the normal fuel, whether a super-abundance of vitamins or analeptics or other substances.

The glutamic-acid story takes a familiar course[1]. The prologue (1936) was Weil-Malherbe's demonstration. In Act I (1946–8) some reasonably careful clinical reports appear, on the basis of which are made claims, backed by reference to maze-learning experiments in rats, that the addition to the diet of 6–24 grammes of glutamic acid per day significantly improves the intellectual functioning of mentally deficient children. These claims are enthusiastically taken up in Act II (1949–51), but towards the end of this Act some discordant opinions are heard. In Act III (1951–55), hopes are dashed by the publication of the negative results of more satisfactory animal experiments and clinical trials. Shortly before the final curtain there is a temporary comeback when the claims are restricted to cases in which there is a history of brain injury.

A belated epilogue, with moral, has now been written by Astin and Ross[2], who summarise their review of the 33 papers published on the topic between 1948 and 1955 in a 2×2 box table ($P > 0.001$).

	Positive Results	*Negative Results*
With controls	6	13
Without controls	14	0

Positive results tend to be obtained when there is no control group. Scrutiny of the six papers in the top left box (positive with control) reveals other flaws which largely invalidate the conclusions, and little or no support is found for the hypothesis that glutamic-acid medication has a specifically beneficial effect on intellectual functioning. It is to be hoped that, when the next hare is started, investigators will not only use control groups but will also take the trouble to develop more suitable tests of intellectual functioning and not rely on intelligence tests devised for quite other purposes.

D. RUSSELL DAVIS

[1] Penrose, L. S. On the Objective Study of Crowd Behaviour. 1952. *London:* Lewis.

[2] Astin, A. W., Ross, S. 'Glutamic acid and human intelligence.' *Psychol. Bull.* 1960, 57, 429–434.

Physical Treatment of Children with Cerebral Palsy

WITH increasing experience comes the knowledge that physical treatment is only one facet of the habilitation of children with cerebral palsy, and one becomes ever more conscious of the roles that are played, *or could be played*, by educationists, psychologists and psychiatrists. Perhaps even more important is the psychological climate in which the child lives and has lived since birth, whether at home or at school and with or without his relatives.

Time and again, when one is constantly associated with cerebral palsy, one is conscious of a retrogression here or a sudden improvement there in the child's physical condition which cannot be explained by the treatment given, but, if one probes more deeply, is found to be associated with the climate in which the psyche lives. (On the other hand, we must beware of too facilely attributing retrogression under therapy to changes in this climate, since it may be due to some other change in the child, or it may be one's own climate that has changed.)

This is the real reason for advocating that education, treatment and psychological care must go hand in hand, that each school or clinic should have an independent co-ordinating authority who can take the wide general view. Obviously this authority should usually be the interested paediatrician, and he should have time to do this part of his work.

Physical Treatment

When we come to physical treatment pure and simple, the whole picture has become bedevilled with 'methods' and personalities. One method is played off against another by its adherents and by over-anxious parents who cannot always be blamed for their attitude. When we consider how little we know of the results of interference with communications between the brain and a functional muscle movement, this situation looks rather silly, and when intolerance creeps in to the extent of advocating this or that method as the 'only' way one's comments must be considerably more harsh.

The physical treatment of cerebral palsy should depend on a careful and continued assessment and re-assessment of the child's functional ability; on the utilisation of *all* our knowledge and ideas, sometimes on an empirical basis and sometimes because one knows how and why; and on the keeping of adequate progress records at intervals of three months, these charts being filled in, not by the therapist alone, but with an independent, interested third party present. It would be useful, if it were possible, to combine a physical progress chart with another 'plot' showing variations in the psychological climate.

No two cases of cerebral palsy have the same physical handicap. Our approach to the minutiae of physical

therapy must therefore be on the widest possible base. We must not let our preconceived ideas prevent us from adopting a set of manoeuvres by way of trial and discarding them if they prove of little use. With experience of the results of treatment, knowledge of other routines, and our own ideas we shall gradually learn the 'know-how' of getting good results, though there will often be children who show very little response.

The Value of Watching the Child

With a new case, the first hours (and not a few of them) should be spent firstly in just watching, and secondly in making complete routine neurological examinations. One notes the movement of the child's limbs, trunk and head, or the lack thereof, in all positions of lying, sitting and, if possible, supported standing. Has he got head control—does he even move his head? Are his limbs stiff or are they hypotonic? Do some muscle groups show increased tension compared with others? Are his trunk muscles weak? Do his limb movements alter with his position? Do his eyes and tongue move normally? These and other questions which will occur to the interested examiner must be asked and answered. As the child gets used to his surroundings and to being handled by his therapist, other queries will arise. Has he a true persistence of the tonic neck reflexes? In spite of expert diagnosis, does he in fact show movements perhaps associated with athetosis in the hands? Or is there some spasticity in his legs though none was suspected at first?

Therapy and Re-education

The purely watching period over, therapy starts in earnest. If there are abnormal persistent tonic reflex patterns, are we to try to inhibit them by posturing or similar manoeuvres? Or are we to attempt to abolish them by getting the child erect on his own two feet as soon as possible? For it must be remembered that in the normal child, by the time he comes to sit erect, these reflexes have become integrated into his total movement complex; too much cannot be read into the behaviour of the decerebrate animal. Are we satisfied that by inhibiting these tonic reflexes, as a result of passive attitudinising, we are going to facilitate a *functional* movement?

If we decide that stretching hypertonic muscle groups with calipers and splints offers the best chance in any given case (see the latest work on abolishing afferent impulses), do we agree that this is the only line to take, or should other ideas be used as well?

Re-education, or perhaps one should say 're-awakening', of afferent impulses may be necessary and should not be omitted. It is well to remember that 'muscle sense' may be absent in trunk muscles as well as the more usual 'joint sense', which may be why some children with a relatively moderate handicap find great difficulty in standing erect. Unaffected muscle groups can sometimes be trained to extend their function to take over from the affected ones, and this is another approach which should be considered. TEMPLE FAY, KABAT and others have ideas which can also be incorporated in any system evolved for a particular child.

We have to remember that in athetosis it is the muscle with the *double function* which so often gives trouble in habilitation.

It is very rare for a child to respond fully to any one idea of physical treatment. A system in which some or even all of the above ideas (not forgetting

surgery) are applied in a co-ordinated whole seems to offer the best chance of progress.

All through physical therapy the effect of the therapy on the child as a person and the child's feelings about the treatment need to be under constant observation and evaluation by the therapist. Is the child physically too tired to profit? Does physiotherapy profit a peevish or an apathetic child? And even if, with a baby, we are not asking for conscious co-operation, does treatment have a good effect if the child cries through most of the session? Is the school term so long that the children get no benefit from the last few weeks? What seems to be needed is a comprehensive utilisation of all suitable ways of treatment, combined with a mobilisation of all that the child's intellect and emotions can give to support the physiotherapy.

J. H. CROSLAND

NOTICE

BACK NUMBERS, BOUND VOLUMES AND THE 'CLINICS'

Nos. 1 and 8 of our first volume, which have been out of print, have now been reprinted, and we can supply any back number, for the time being, at 5*s*. a copy, post free. The reprinting also allows us to offer more bound volumes. Copies of Volumes 1 or 2, bound in scarlet cloth with index, will shortly be available, while stocks last, at £1, or $3, also post free. As before, subscribers can have their own copies bound free of charge.

The charge for single copies of the *Little Club Clinics in Developmental Medicine* will be 10*s*. or $2, post free.

E. Clayton-Jones

Cerebral Palsy in South Africa

Two surveys of South African children and adults with cerebral palsy were done by Dr. C. H. de C. Murray for the National Bureau of Educational and Social Research in 1957–1958, with the primary object of providing information which would help in planning the care of adult cases. This objective has largely been achieved. The substantial amount of carefully analysed data presented in Dr. Murray's report* must be of great and continuing value to the Cerebral Palsy Division of the National Council for the Care of Cripples in South Africa, in making their plans for the future. It was estimated that at least two-thirds of the 380 children in the major survey would require some form of custodial care on leaving school, and the probability that this rather gloomy prognosis was accurate was increased by the data from the subsidiary survey of adults, for only a third of 111 adults with cerebral palsy were employed outside their homes. The children, however, were a selected group, and many mildly handicapped children able to go to ordinary schools were excluded.

The major survey included children of European descent at certain special schools in South Africa and a few children not at school. Their ages ranged from 1 to 21 years, with 'a few cases older than 21 years'. The intelligence of the majority was found to be below normal, where 'normal' indicates an I.Q. of 80 or above, and generally speaking there was a direct relationship between the severity of the mental retardation and of the physical handicap. For example, among the 189 children whose intelligence was tested according to the standardised South African Individual Scales, 38 per cent of those with mild degrees of physical handicap had I.Q.'s of 90 or higher, compared with 31 per cent of those moderately handicapped and only 15 per cent of the severely handicapped group. The data on personality traits were of particular interest, because estimates of physical and mental handicap alone often do not give an accurate indication of the future response to treatment or of future employability, both of which depend also on personality. In the survey, the children were assessed according to their disposition, their sociability, their persistence, and their ability to concentrate. The results indicated that most of the children displayed the positive quality of sociability, and that a majority were lacking in persistence. The trends in the attributes of ability to concentrate and disposition were not statistically significant, although the results did suggest that the children have difficulty in concentrating, and this was especially apparent in children under 6 years of age. The ratio of males to females was 3:2, the degree of physical handicap was about the same in the two sexes, and about equal proportions spoke primarily Afrikaans and English.

The practical usefulness of these

* A Survey of the Physical and Mental Status of Cerebral Palsied European Children at School in the Union of South Africa. A Survey of the Adult with Cerebral Palsy. By C. H. de C. Murray. Pretoria: National Bureau of Educational and Social Research, 1959.

surveys within the Union of South Africa is unquestioned, but a number of features detract seriously from their general scientific value. The studies were conducted by questionnaire, and the cases were not personally assessed by one experienced doctor. The diagnosis of cerebral palsy, and the assessment of the type and severity of the motor handicap, were thus made by a large number of individuals of varying experience and enthusiasm, and although an attempt was made to standardise criteria by issuing a schedule of instructions, the reliability of results obtained in this way must inevitably be suspect. For example, one doctor diagnosed rigidity in 11 out of his 23 cases of cerebral palsy, and although in this instance the children were reviewed later by a specialist and the diagnoses changed, the incident does not increase confidence in the other returns. This difficulty is indeed acknowledged by the author, but the fact that he rejected 'a few questionnaires . . . about children who were not cerebral palsied, e.g. microcephaly' does not entirely answer the objection. Furthermore, it is not customary in most countries to include brain-injured children without motor involvement as cases of cerebral palsy, and although this is apparently justified in South Africa by the fact that 'some cerebral palsied children with motor involvement show the syndrome of brain injury . . .', it would surely have been more logical to exclude this small number of cases (17 out of the total of 380 cases) or at least to differentiate them clearly from the rest.

The results of these two surveys have been subjected to very careful statistical analysis by the author, but it is open to question whether such precise methods are properly applicable to data of such doubtful accuracy, and they may indeed mislead by suggesting to the reader that the original observations were more reliable than in fact they were.

We are told in the foreword that 'the way is now open for a comparison with findings in other countries'. Alas! the way is far from open. No comparisons of prevalence of the different types of cerebral palsy in the community can be made, for the cases in these two surveys were highly selected groups and many affected children were not included, for a variety of reasons. The majority of the children were in special schools, but the group is apparently not even representative of children in such schools for the institutions were self-selected by their willingness to co-operate and no details are given of those which replied unfavourably. Certain comparisons can of course be made between groups of highly selected cases, although their value is always somewhat limited by possible variations in diagnostic criteria in different countries. Where the interpretation of criteria *within* individual surveys is as variable as it is likely to have been in these, however, the usefulness of comparisons of such surveys with those from other countries is very greatly reduced. Moreover, the implications of statistically significant differences are not easy to assess in such highly selected material.

In fairness to Dr. Murray, it must be said that he recognises many of the difficulties mentioned here, which are indeed those encountered by anyone attempting to carry out a large-scale survey. The purpose of such a survey must be very carefully defined before it is undertaken, for the expenditure in time and money and the requirements for skilled workers must be balanced against the probable value of the information it is expected to yield. For planning its future activities, the

National Council for the Care of Cripples in South Africa would not require a very high degree of scientific accuracy, and the merits of these two surveys must be judged against the background of the expenditure involved and the results desired. By these standards the surveys must be considered useful and worthwhile contributions, and likely to be of great help to workers in the field of cerebral palsy in South Africa. ROSS MITCHELL

Letters and Questions Please

IN this issue the *Bulletin* has acquired a new section—*Letters to the Editor*—and also what will perhaps become a regular feature: *What's to be Done?* Contributions to either will be welcomed.

In a two-monthly journal correspondence cannot aspire to the rapid turnover of *The Times* or even the topicality of *The Lancet* or the *B.M.J.* But, as the first examples of *Letters to the Editor* demonstrate, it can provide an opportunity for putting forward new ideas that others will comment on, criticise and possibly carry a stage further, and for the well-considered discussion of points made in original articles, editorials or elsewhere. The two-monthly has this advantage over the weeklies and dailies—that, with the Editor's connivance, questions can be both raised and answered in one issue, thus offsetting some of the disadvantages of infrequent publication. Letters for publication should therefore be sent as soon rather than as late as possible. In everyone's interest, letters should be economical in verbiage and crystal-clear, for they will be read by many people not wholly familiar with the English idiom.

For *What's to be Done?* the first necessity is a steady flow of arresting questions likely to draw forth views and information of wide application—puzzling cases, tricky technical points, experiences on which others may throw light—there must be plenty of these in our readers' everyday work to keep our new feature well supplied with suitable questions. The chosen ones will be submitted to likely respondents, with results that may well prove interesting as well as instructive.

THE EDITORS

The Subacute Encephalitides

PROF. LUDO VAN BOGAERT, M.D.
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A sequel to the paper on acute encephalitides in *Bulletin* Vol. 2, No. 3, 1960, pp. 131-134.

THE subacute encephalitides have been seen in Western Europe sporadically since the epidemic of encephalitis lethargica in 1923-25. At that time they were described as atypical variants of this disease. Dawson (1933) demonstrated the presence of an unusual number of acidophil intranuclear inclusions in the subacute encephalitides. These inclusions had special staining properties and cytological characteristics which, he thought, made it possible to differentiate them from those described in encephalitis lethargica.

It was not until 1938 that in various places attention was paid to the subacute encephalitides, and the question was raised whether the atypical cases were not related to the Japanese and American encephalitides, called 'Type B encephalitis', in contra-distinction to encephalitis lethargica or von Economo's disease, also called 'Type A encephalitis'.

Thus these atypical cases have been described successively as inclusion body encephalitis (Dawson 1933), and 'atypical nodular encephalitis' similar to encephalitis Japonica (Pette and Döring 1939). The protracted course of Dawson's cases makes it difficult to speak of them as subacute encephalitis; in Pette and Döring's series, however, some cases have a shorter evolution and can certainly be classified as acute encephalitides.

In 1938 we made a different approach to the study of these same diseases. At that time our main interest was to break up the group of diffuse sclerosis, and more particularly to isolate from that group a so-called inflammatory form, so that in our first examination we were especially investigating the nature of the lesions in the white matter. When with J. De Busscher (1939) we observed our first case of subacute encephalitis, we were particularly anxious to differentiate this from the diffuse inflammatory multilocular sclerosis, which some called inflammatory.

Now that our personal experience is based on some 80 morphologically verified cases, it gradually becomes clear that inclusion body encephalitis, nodular panencephalitis and subacute sclerosing leucoencephalitis are one and the same disease. At present, according to whether the histologist has been impressed by the inclusion bodies, the inflammatory localisation in the grey matter, or the inflammatory infiltrations in the white matter, one is tempted to classify a given case under one of these three terms.

The different appearances are neither for nor against an identical aetiology. It would, however, be equally valid to say that each of these tissue responses is caused by a different agent as to favour the hypothesis that the different responses are the expression of an individual neuronal reaction based on certain characteristics of the subject's constitution.

This aetiological problem is not going to be solved by our clinical or morphological methods, but whatever name is applied to these subacute encephalitides, their symptomatology, clinical evolution and histopathological picture are identical and make it possible to recognise the disease, which is the important point.

Subacute encephalitis does not arouse the interest of either the neurologist or the orthopaedic surgeon concerned with the treatment and diagnosis of cerebral palsies. The evolution of the disease is usually so

rapid that the symptoms do not have time to become static, and the abnormal postures that one can observe have too changeable a pattern to be suitable for instrumental or physiotherapeutic correction.

These affections should, however, concern the paediatrician interested in neurology or psychiatry, because they manifest themselves by intellectual and personality changes and educational difficulties. The condition produces a hyperkinetic picture with epileptoid features which may lead the unwary observer to diagnose some rare disease.

The cases that develop slowly and remain at a stage in which the manifestations of the disease are psychogenic are of particular educational and medical interest. We and our collaborators have stressed this point from the beginning. Even today these psychogenic manifestations are less well known than the terminal neurological picture. That is why we want to recall them. In this paper we will separate them from the disintegration of the nervous system.

Stages of Mental Involution

In the *first stage* of the disease one is impressed by the deterioration in the patient's mental state. This begins insidiously and manifests itself chiefly by behaviour disturbances. The child presents a psychomotor instability, and the schoolteacher notes difficulties in attention and concentration at work. The duration of this period varies in different cases. These symptoms rarely attract attention and the parents ultimately consult the physician because of the child's epileptic seizures or the 'absences' which accompany the symptoms of incipient dementia.

These disturbances of attention can be seen in children who are merely nervous or maladapted. In the latter condition, however, the parents will say that the symptoms have been present from early childhood, whereas the symptoms of subacute encephalitis appear in children whose psychological development has previously been normal. One sees model children become undisciplined, irritable, hyperemotional, and hypersensitive; their mood is variable, at one moment they are querulous, then aggressive, then naughty, they become more rude and more teasing, and very soon their school-work deteriorates. This mixture of instability and anger, disobedience and indiscipline may well be lacking, and then the child becomes insidiously more indifferent and passive, thinking is retarded, initiative is lost, and the child is no longer interested in his own or his companions' games. These states may alternate.

This phase usually lasts a few months but may cover a longer period, and it can almost always be detected by careful questioning of the family or the child's teacher.

In the *second stage* this regression becomes more general—attention and concentration are seriously impaired; the formation of ideas is slow, fatiguability is extreme, verbal expression becomes incoordinated, and one sees states of confusion which may last several minutes. When speaking to these children one sometimes gets the impression that one is dealing with dementia, but next day one notices that some mental functions are still intact; writing, arithmetic, speaking, reading and gesticulatory expression disappear slowly. The impairment of memory is obvious. There may be signs of aphasia and apraxia. At this stage one may observe a disassociation between volitional and automatic reactions. The child is still capable of writing his name or of copying, but spontaneous writing is no longer possible. The little patient deforms sentences and words. Gradually he can only read short sentences, certain words or syllables, and no longer understands what he is reading. He becomes incapable of dressing himself, disorientated in space and confused; finally, all higher functions of the nervous system disintegrate.

In the *terminal stage* of the disease the dementia is profound. The child retains a certain affective relationship with his environment and family, but all voluntary activity disappears. The child sinks into a purely vegetative state.

The Clinical Picture

When one considers the general picture of these disorders, it is clear that some characteristics of this mental state are common to all forms of mental deterioration and link this leucoencephalitic dementia with the group of evolutive dementias. However, as one of our collaborators has emphasised, this involution has some distinguishing characteristics. Its onset is characterised by insidious and non-specific personality changes; the dementia takes different forms according to the child's age when the disorder begins. At puberty the dementia is characterised by the same personality changes as are seen in dementia praecox, but symptoms of cortical deficiency are more prominent. At school-age, on the other hand, the personality changes are not so obvious, and whereas before school-age aphasia predominates, now the dementia particularly affects knowledge acquired at school, and it develops without passing through a stage of automatico-voluntary dissociation. The last important fact which should be stressed is that the dementia is total and progressive and affects all mental functions.

Neurological symptoms accompany each of these periods of mental involution (Figs. 1, 2 and 3).

The first period is characterised by falls. These in fact represent generalised myoclonic jerks. The child falls at the slightest provocation or spontaneously; the fall is of a short duration without any subsequent symptoms. Despite warnings from the bystanders these falls become more frequent and are accompanied by some disturbance of consciousness which finally lasts long enough to attract attention. These sudden falls are described either as jerks or as loss of tone. If one watches them closely one is struck by their periodicity, which parents usually notice at an early stage.

Fig. 1A.

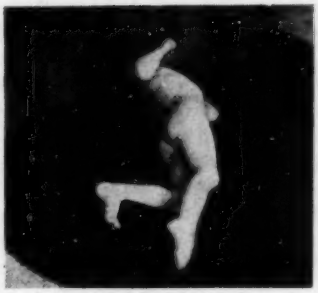

Fig. 1B.

Fig. 1. (A) Involuntary movements of the upper limbs with thrusting jerks of the right leg and opistothonos.
(B) Torsion of the body.

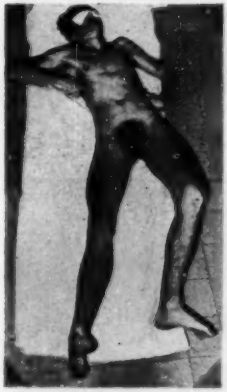

Fig. 2A

Fig. 2B

Fig. 2. (A) Note the hyperextension of the right leg, the pronation and hyperflexion of the left hand, the opistothonos and the peculiar posture of the right hand with the fingers hyperextended.
(B) When the patient is standing supported, note the hyperpronation of the right arm with extension of the index, and the hyperlordosis. (Case observed by van Wymeersch, Macken and Guazzi.)

In the second period, when there is a progressive deterioration of mental function, a whole range of hyperkinetic disorders appear, from myoclonic jerks to choreothetosis and hemiballismus; convulsions occur which may be either grand mal, petit mal or Jacksonian in type. Once again, all these hyperkinetic manifestations show a periodicity which usually remains constant in the same case, variations occurring only occasionally.

Fig. 3A

Fig. 3B

Fig. 3C

Fig. 3D

Fig. 3E

Fig. 3F

Fig. 3. (A) Opistothonos with forced opening of the jaws and involuntary opening of the mouth.
(B) Rotation of head to the left.
(C) Onset of ballistic movements of the right upper limbs, with hyperextension of the fingers.
(D) The movement is completed by an involuntary elevation of the elbow, and an accentuation of the hyperpronation of the forearm which results in a complete overturning of the hand.
(E) Returning of the hand to its initial position with forced hyperextension of the head.
(F) Same movement of the left hand but in hyperextension and accompanied by a sudden flexion of the two lower limbs.
(Photographs taken in the terminal stage.)

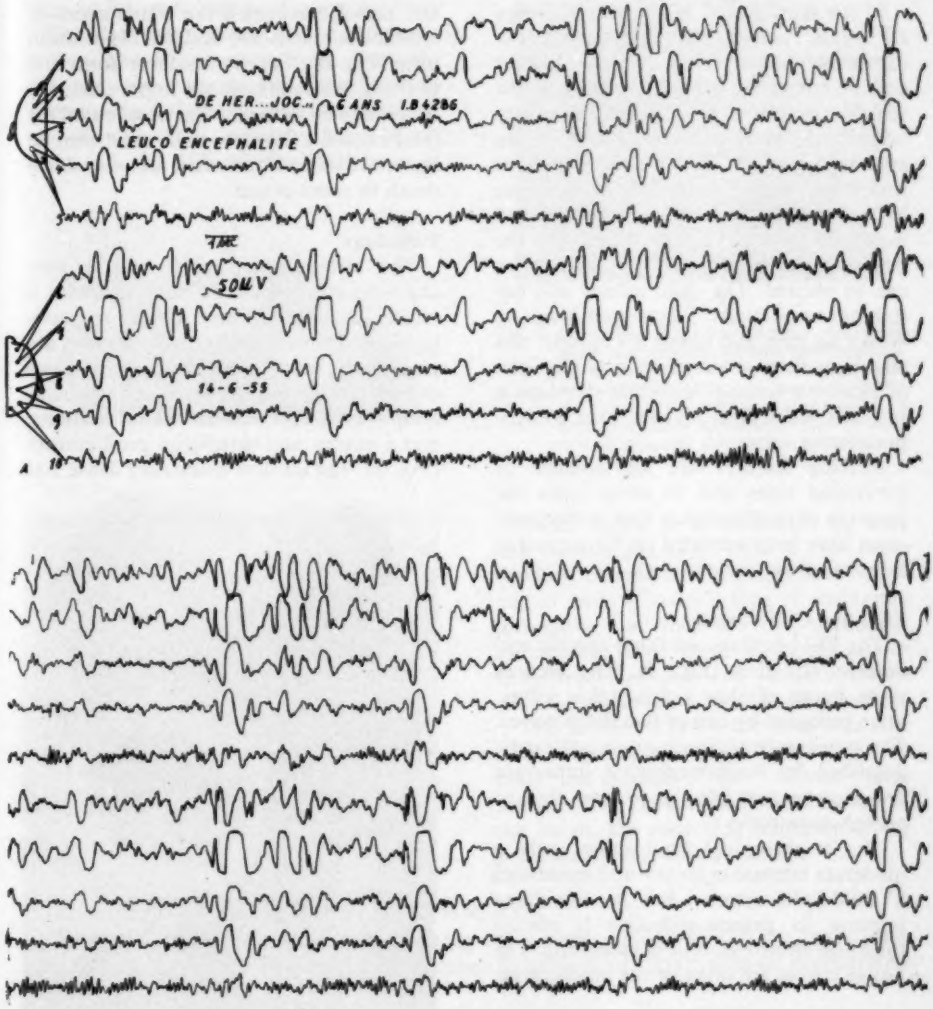

Fig. 4

Fig. 4. Typical electroencephalographic patterns of subacute sclerosing leucoencephalitis. Necessary criteria for the diagnosis:
1. Presence of slow paroxysms regularly repeated during the whole record, even if it is extended for an hour or more.
2. The frequency of this regular succession of paroxysmal bursts is stable in a given EEG (middle values 6–12 bursts per minute, the extreme values recorded being from 3–4 to 15–18 bursts per minute).
3. The morphology of the bursts remains on the whole almost the same in a given derivation, but it varies generally from one derivation to another.
4. These abnormalities are always bilateral, and almost always symmetrical and synchronous over the two hemispheres.
(NOTE: The lower half of this figure is a continuation of the upper half).

In the third period of the disease, when all mental activity is disappearing, the involuntary movements become scarcer and are not strong enough to overcome the spastic paraplegia or quadriplegia. Only occasionally does one see an arm or leg assume a peculiar posture. The body is sometimes hyperflexed and sometimes hyperextended, the face wears a contracted mask, the jaws are tightly closed and the fists clenched. Occasionally a grasp reflex can be elicited. The child groans and has sighing respirations*; now and then he blinks his eyes and opens his mouth; this also may show some periodicity. In spite of intensive feeding, the child develops a cachexia which precedes a terminal broncopneumonia.

Striking features are the absence of pyramidal signs and in some cases the presence of papilloedema. This is why some cases have been operated on for suspected tumour of the parietal area.

EEG and CSF Findings

The EEG findings are fairly specific and deserve attention (Fig. 4). The records show bursts of slow polymorphic waves, often preceded by one or two sharp waves. The appearance of these paroxysmal bursts, described by Radermecker, is important and they are now familiar to most electroencephalographers.

The cerebrospinal fluid shows only a moderate increase in protein and sometimes a mild pleiocytosis, but a significant increase in gamma-globulins is always found on electrophoretic examination. The Lange curves are often of the paretic type.

Aetiology and Treatment

The cause of subacute leucoencephalitis is still unknown. Some think that it is an atypical and subacute form of encephalitis due to herpes simplex virus, but up to now there has been no proof of this view.

Unfortunately there is no effective treatment for these subacute encephalitides and the outcome is always fatal. My co-workers and myself have tried the whole series of antibiotics, cortisone, and ACTH without observing any benefit whatever from any of them. We even have the impression that in some cases cortisone has accelerated the fatal outcome. Symptomatic treatment of the convulsions is indicated and may delay death to some extent.

Pathology

The histopathological picture is very characteristic indeed. In some respects it recalls juvenile GPI and African trypanosomiasis. Primary infiltrative processes are particularly marked in the subcortical parts of the white matter, comprising perivascular lymphocytic and plasma-cell infiltrations, and a micro- and astro-glial proliferation (Fig. 5). The cortical lesions are dense but

Fig. 5

Fig. 5. General view of the cortical and subcortical lesions: discrete meningeal reaction, disseminated cortical lesions, consisting of acute cellular injury, pericapillary infiltration and microgliosis. Note the increase in perivascular cuffing in the deep layers IV to VI and in the subcortical areas dense gliosis of the white matter. (Celloidin, Cresyl violet.) (Case observed by Macken and Lhermitte 1950.)

* 'The windy suspirations of forced breath.'—*Hamlet.*

vary in intensity, comprising pericapillary infiltration, acute neuronal and ischaemic lesions, and diffuse microgliosis. Demyelination is also variable, but almost always present; it is diffuse, looks like worn fabric, and shows marked and early fibrous gliosis (Figs. 6A and 6B). Myelin breakdown of

Fig. 6A

Fig. 6B

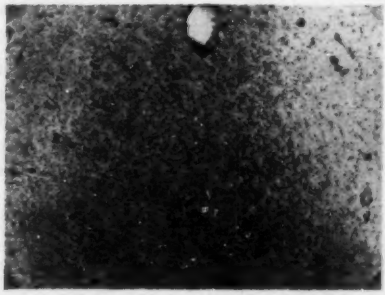

Fig. 6C

Fig. 6. (A) Diffuse demyelinated areas, resembling worn fabric (frozen section, Spielmeyer). (B) Fibrous gliosis in a corresponding section (frozen section, Holzer's technique). (C) Myelin degeneration surrounded by microglia (fluffy appearance) and by compound granular corpuscles (large round structures). (Cresyl violet, Scharlach R.)

variable degree is accompanied by fixed and mobile glia (Fig. 6C). If one looks carefully one can find in a significant number of cases Cowdzy's type A eosinophil inclusions, but they are not pathognomonic; one finds them in encephalitides of different aetiology and we do not consider them characteristic of these disorders.

This is why we prefer the term 'subacute sclerosing leucoencephalitis' to 'inclusion body encephalitis'. The former term indicates the predominant distribution of the lesion, the sclerosing tendency of the process, and its subacute course.

SUMMARY

The subacute encephalitides are becoming increasingly common, and the cases often pass unrecognised because of their resemblance to unadapted extrapyramidal degenerative disease.

The various forms of the condition and their symptoms are described.

Unfortunately the treatment is still wholly ineffective and the cause remains unknown.

RÉSUMÉ

Les Encéphalites Subaiguës

Les encéphalites subaigués deviennent de plus en plus fréquentes. Les diagnostics ne sont souvent pas précisés à cause de leur ressemblance avec les maladies extrapyramidales dégénératives.

Les différentes formes et leur sémiologie sont décrites.

Malheureusement le traitement est encore totalement inefficace et la cause en demeure inconnue.

ZUSAMMENFASSUNG

Subakute Encephalitides

Die subakuten Encephalitides werden immer häufiger und die Fälle verlaufen oft, ohne erkannt zu werden, weil sie den extrapyramidalen Entartungskrankheiten ähnlich sehen.

Die verschiedenen Formen der Krankheit und ihre Semiotik werden beschrieben.

Leider ist die Behandlung noch völlig unwirksam und die Ursache unbekannt.

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Piaget's Approach to Problems of Intellectual Development

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THE work of Jean Piaget has recently been attracting increasing attention in the English-speaking world. His contributions to Psychology in general, as well as to Child Psychology and the Philosophy of Science are so immense and revolutionary that it is beyond the scope of any single article to do more than touch on certain of his basic ideas. Although a number of authors have in the past attempted to give general descriptions of his system of thought, we shall attempt no more than to orientate the reader to the way in which Piaget approaches the problem of intellectual development in children.

For several decades the customary approach in this country to the problem of assessing the intellectual development of the child has been a *psychometric* one; the child's level of intellectual functioning is evaluated against a background of average achievements in a number of tests at various age-levels. Thus a child of 7 years who can perform successfully on a set of tests located statistically at age 8 is thought of as possessing an intelligence somewhat superior to that of the average child of his own age. Piaget's concern is a completely different one. For any intellectual achievement of the child, he will ask: How did this come about? What concepts and processes are involved? How does the child's achievement compare with an apparently similar achievement in an adult?

Two Examples

This difference of approach may be illustrated by citing two examples taken from the Terman-Merrill revision of the Stanford-Binet tests of intelligence.

Example 1. One of the tests located at age 5 years in Form L of the Terman-Merrill revision is 'copying a square'. In this test the child is presented with a figure of a square and is asked to 'make one just like this'. In their discussion of this particular test Terman and Merrill suggest that successful performance depends on an appreciation of spatial relationships, and the ability to make use of visual perceptions to guide a rather complex set of motor co-ordinations.

For Piaget the ability to copy a square is only a single link in the developmental chain of mastery of spatial representation in the child. At birth the child has no notion of space at all. Indeed, the very ideas of space—of there being *a* space instead of discrete sensations representing a multitude of buccal, tactile-kinaesthetic, auditory, visual and motor experiences—does not exist until later. In the earliest weeks there is not even one body-space but

each experience of the body represents momentary bodily sensations only. Slowly, with increasing sensori-motor experiences, sensations become associated; for example, the child gradually learns to turn his head in the direction of a noise. The child has to discover that when he moves his legs this movement occurs in the same space as movements of his head, and that head and legs form only part of the body as a whole. Ultimately the body is discovered to be only one thing in a space containing other objects, which are differentiated and endure in time. This complex development takes place during the whole of the sensori-motor phase, which according to Piaget covers approximately the first 18 months of life. In this phase, the role of motility is all-important, as the fundamental spatial links cannot be made without repeated physical manipulation of the environment and the co-ordination of changing experiences which results from the child's activities. For Piaget, physical activity is vital for the development of all aspects of intelligence; even the most complicated of thought processes represents internalised activity.

By the time the child is 18 months old, he will be able to move around his relatively limited world, and will have a perception of space quite adequate to his needs. But he will as yet have little capacity for spatial representation as distinct from his ability to perceive things in space. To achieve spatial representation he will have to move on to a further level of development, and to discover his world once again in terms of newly-gained co-ordinations. He will have to progress once more from an egocentric point of view (that is, one closely linked with his own subjective experiences and activities) to a more objective view of the world.

One facet of the development of spatial representation in the child is the growth of the capacity to copy figures. Piaget has performed an immense number of experiments to trace the growth of spatial representation in the child, and of these some relate to the drawing of simple geometrical forms. The study of children's attempts to copy such simple forms has demonstrated that, although these forms may be immediately recognised and identified, the process of representing them with pencil and paper involved, at certain ages, some difficulties. Some figures could be copied easily by children of 3–4 years, while they failed to copy others. Those which could be duplicated involve certain 'topological' relationships—they necessitate only a 'functional' geometry which preserves such relations as 'under', 'over', 'between', 'outside', and so on, rather than the exact relationships of size and angle characteristic of Euclidean geometry. Thus children of 3 years, when presented with, say, four shapes—a triangle, a square, a closed irregular shape with a dot inside it, and a line with a dot over it—could easily copy the last two but were unable to copy the triangle and square. The figures which they could copy involve only the notions of 'closedness' and 'interruptedness' and of 'inside' and 'above'. With no idea of angular representation, the child of 3 regards a triangle, a square and an irregular closed figure as having the same geometrical properties. Topological or functional representation is closer to the child's actual behavioural activities than Euclidean relationships which necessitate more complicated co-ordination.

The child who has the ability to draw a square has reached a stage in the development of his capacity for spatial representation which follows well after the ability to perceive squares as such, and after the capacity for topological representation has been achieved. He has taken one step along the path towards complete spatial representation, and will only later be able to represent more complex figures in space. Thus it will take another two years for the child to be able to copy a diamond-shaped figure (located by Terman and Merrill at age 7 years). This can only be achieved when the child possesses the ability to refer his drawing to a geometrical frame of reference which he carries in his mind and which is readily available to guide him in

his progressive representation of the figures. No such frame of reference is necessary for copying a square, where the child has only to represent the relationships between two adjoining lines, and can build up the figure on the basis of these partial relationships.*

Example 2. At year 6 in Form M of the Terman-Merrill revision a test of 'number concepts' is presented. Briefly the test is as follows: The tester has twelve cubes arranged in a pile in front of him. The experimenter asks the child to place two blocks on the paper. The blocks are then restored to the pile, and the child is asked to place ten, three and six blocks on the paper in the same way. If three of the four attempts are correct, the child gains credit for the test, and is assumed to have a concept of 'number'.

Such a test would appear, to all intents and purposes, to indicate the child's possession of numerical concepts, but Piaget has found it necessary to probe more deeply. He has pointed out that the ability to co-ordinate the activity of counting out a certain number of cubes, with the appropriate verbal numbers, by no means implies that the child possesses a concept of 'number' as such. For a child to have such a concept he must be able, for example, to abstract the numerical properties of the objects he is examining, taking no account of such factors as their size, their arrangement in space, or any other of their qualitative properties.

Let us consider a child who is confronted with the following situation, one of the many experiments performed by Piaget in his investigations of the number concept. The tester explains to the child that he has a bag of counters, and that he is going to place these singly in a line in front of the child. The child is asked to copy the tester as he places each counter. When the tester has placed ten counters on the board, and the child has placed his ten correspondingly, the tester asks: 'Have we the same number, or has one of us more or less than the other?' The child may answer unhesitatingly that both have the same number of counters. The tester now puts his ten counters in one pile, and divides the child's into three separate piles. He then asks: 'Do we now have the same number of counters, or has one of us more or less than the other?' Before the age of 5½ or 6 years the usual answer will be: 'Oh, but I have more than you, I have three piles.'

This experiment shows that a child of 5 years may be misled in his judgment of number by the immediate visual impact of the arrangement of the counters. The grouping into three piles gives an impression of 'more than one' pile, and the child's notion of number has not yet reached a level of conceptualisation sufficient for him to disregard his immediate and intuitive impression. If the child, in this particular experiment, is encouraged to examine his counters more closely, to rearrange them opposite the tester's in their original position (in short, to reverse the physical activity of moving the counters) he might well correct his previous answer and say: 'I was mistaken; of course we each have the same number.' However, if subsequently the conditions of the experiment are changed, he might again fall into his original error. Such a 5-year-old child, who has not yet a concept of number in the Piagetian sense, might well, if his verbal abilities are above average, be able to pass a test of 'counting' such as the Terman-Merrill 'number concept' test described earlier.

What Piaget regards as essential in the attainment of a concept of number, as distinct from the ability to count, is the possession of the capacity for manipulating the counters *mentally*. The child has to detach his thought processes from what he perceives, has to be able to think of all sorts of possible transformations of the material, and must be able to *reverse* a

* An interesting experiment of Piaget's neatly illustrates this point. A child of 6 years who cannot copy a diamond may be able to do so when the tester joins two corners of the figure to form two adjacent triangles.

train of thought, so that he again reaches his original point of departure. The essence of what Piaget calls the 'reversibility' of thought processes is that they are liberated from the influence of immediate perception, and reach the level of logical thinking. Logical thought, for Piaget, consists in the internalisation of activities, and the correlated development of abstract frames of reference of an operational kind. No real concept of number is possible unless this level has been reached.

SUMMARY

In an attempt to demonstrate the way in which Jean Piaget looks at intelligence and its development, a comparison was made with the more usual psychometric approach. Two subtests of the Terman-Merrill revision of the Stanford Binet intelligence test were taken as a basis for this comparison, and it was shown that Piaget's interests in this field are directed towards the development of the thought processes which lie behind apparently simple tasks. He is not primarily concerned with the allocation of abilities to various age levels, but rather with the development of the internalised structures and mechanisms which enable the child to understand and solve various intellectual problems.

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Problems of Hypoxia in the Genesis of Cerebral Palsy

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THE pathology of the brain in cases of cerebral palsy is very varied and may involve both cortical and subcortical structures. Some important examples of the former have been surveyed in this *Bulletin* by Crome (1959), who has also pointed out how difficult it may be to decide the various factors responsible for a particular pathological end result.

Among the cases whose origin is believed to be para- or post-natal the causative factor of hypoxia is frequently encountered. By this is meant some reduction in the oxygen supply to the nerve cells of the foetal or infantile brain which may be brought about in several ways, including a reduced oxygen saturation of the blood, a reduced blood-flow (ischaemia) to the brain, or a consumption of oxygen by nerve cells in excess of the available supply.

Production of Hypoxia

Neonatal asphyxia is perhaps the most obvious situation resulting in cerebral hypoxia and is regarded as a major cause of cerebral palsy (Courville 1953). It is interesting to note that the distribution of lesions in such cases reveals a pattern of selectively vulnerable structures including the cerebral cortex (particularly its third layer), amygdaloid nucleus, thalamus, Ammon's horn (or hippocampus), corpus striatum and cerebellum. It is the recognition of this pattern of lesions in the brain that leads the pathologist to infer the occurrence of some episode of hypoxia.

Similar effects may result in the foetal brain if the oxygen-carrying capacity of the maternal blood is reduced by a severe anaemia and will also occur if placental function is disturbed, as in Caesarean section performed for placenta praevia.

Many cases of cerebral palsy can be ascribed to 'birth injury', yet mechanical factors alone cannot explain the ultimate state of the brain in every case. Thus excessive moulding of the foetal head may produce tears in the edge of the tentorium with consequent rupture of veins or venous sinuses. The escaping blood, whether inside the brain or on its surface, can act as a space-occupying lesion and by displacing brain tissue may interfere with its blood-supply. Thus an ischaemic factor is introduced in addition to the original mechanical one.

Epilepsy is a symptom that features frequently in the case-histories of children with cerebral palsy. It may follow closely on an abnormal birth or occur later as a complication of even a minor head injury or some childhood infection (Meyer, Beck and Shepherd 1955). Epileptic seizures are of pathological significance because, when severe and frequent (including status epilepticus), they can produce further damage in an already abnormal brain. Here again, the superadded lesions are found to occur in the vulnerable areas already listed, suggesting that epilepsy damages the brain by producing cerebral hypoxia.

The intensity of hypoxic lesions may vary widely from case to case, but there is in the human brain no essential difference between the pathological effects of hypoxia

and of ischaemia. Reduced to the simplest terms, hypoxia of any type will exert its effects on both the neurone and the cerebral capillary. In the latter, damage to the endothelial cells leads to oedema, haemoconcentration and stasis. However, in order to explain the pattern of selective vulnerability to hypoxia, additional factors must be sought.

Localising Factors

Originally, two major hypotheses were advanced to explain the pathological findings. The 'vascular' hypothesis of Spielmeyer (1925) and his school regarded vasomotor changes, including vasospasm, as the major factor. The evidence for such vasospasm has been regarded as unconvincing by Meyer (1958), based as it was on postmortem histological examination. The second hypothesis, that of C. and O. Vogt (1922), attributed the vulnerability of a region to certain physiochemical (pathoclitic) properties which rendered it susceptible to hypoxic damage.

More recently, Scholz (1952) has combined the vascular and pathoclitic hypotheses by suggesting that the pathoclitic properties of a particular structure render it vulnerable to hypoxia but structural damage is finally brought about by the addition of vasomotor disturbances widely distributed in the brain.

A further localising factor is thought to be the level of functional activity of cells or cell groups. Thus, under conditions of general hypoxia, an active centre, by virtue of its greater oxygen utilisation, might be expected to show evidence of injury before a less active one. This factor of 'consumptive anoxia' (Scholz 1952) has been advanced to explain the typical lesion in the Ammon's horn in epilepsy when the level of its electrical activity is known to be high. The experiments of Meyer and Portnoy (1959) have shown (in the monkey) that during an epileptic seizure there is, in spite of a rise in blood-pressure and in cortical blood-flow, a dramatic fall in cortical oxygen tension.

Vascular Compression

At the moment neuropathological attention centres on mechanical vascular rather than upon intrinsic neuronal factors. This approach dates back to Adolf Meyer (1920) who explained infarction of the occipital lobe in a case of secondary brain tumour as a result of compression of the calcarine branches of the posterior cerebral artery against the tentorial margin. Later workers have endorsed this view, while recently Lindenberg (1955) has suggested that similar compression can occur in the internal carotid, anterior choroidal, superior cerebellar, posterior inferior cerebellar and other smaller arteries. Lindenberg believes that this mechanism can account, not only for the sclerosis of the Ammon's horn, but for all the other lesions comprising the pattern of selective vulnerability in states of hypoxia.

It must be pointed out that hypoxic states cannot be regarded as comparable to acute space-occupying or traumatic lesions, which form the bulk of Lindenberg's material. They differ in that there is a lesser rise in intracranial pressure, tentorial herniation is less marked or even absent, and the brain-stem haemorrhages so often associated with major internal herniae are hardly ever seen.

Further, the hypothesis of vascular compression involves the assumption that intracranial pressure can exceed systolic arterial blood-pressure, although manometric proof of this situation is lacking in man. Norman and others (1957) believe that this is likely to occur only when there is some systemic hypotension such as may occur in neonatal shock. Apart from this latter situation there is no convincing evidence that the rise in intracranial pressure due to hypoxia is coincident with a fall in blood-pressure. This applies particularly to the epileptic seizure. Thus Small and Woolf (1957) have described severe anoxic lesions in the brain of an epileptic without any rise in intracranial pressure or tentorial herniation. Scholz (1959) has also stressed the absence of brain-stem lesions in the pattern of ictal damage. It would also seem

reasonable to suggest that, within the vascular bed, veins are more likely to suffer occlusion than arteries. The production of infarction as a result of a sudden rise in venous pressure has been demonstrated experimentally by Denny Brown *et al.* (1956).

It must be concluded that, at the present time, the hypothesis of arterial compression as the cause of lesions in hypoxic states '. . . still needs confirmation and in particular experimental proof'. (Gastaut *et al.* 1959). It appears to rest on a no more secure basis of fact than the earlier hypothesis of vasospasm, and this latter, in my view, equally merits investigation by experimental techniques.

I am not suggesting, however, that vascular occlusion in some form and at some site between artery and capillary is to be dismissed as improbable in hypoxia. Burton (1951), in defining the 'critical closing pressure' in arteries and arterioles, has indicated some of the conditions under which vascular occlusion can occur. It appears that hypotension of a degree sufficient to attain the 'critical closing pressure' will arrest blood-flow not at an arterial but at an arteriolar or capillary level. There seems to be an urgent need for the application of this approach to the cerebral circulation during experimental hypoxia, providing that the basic variables of intracranial pressure (above and below the tentorium) and systemic blood-pressure can be simultaneously recorded.

Finally, it should be remembered that it is in the human brain that purely anatomical variables will attain their maximum importance, and of these, variations in the size and shape of the tentorial hiatus (Corsellis 1958) and in the size, symmetry, calibre and course of the vessels of the circle of Willis will be of cardinal importance.

Discussion

The various hypotheses of the pathogenesis of hypoxic lesions that I have briefly outlined indicate, above all, the difficulties inherent in the neuropathological approach, based as it must always be on a retrospective interpretation of events and almost always in the absence of critical physiological information.

It is most probable that while each hypothesis can contribute to the explanation of some of the phenomena, no single hypothesis can explain all. Strong support for the factor of 'consumptive anoxia' has come from the experiments of Meyer and Portnoy, but there have been few investigations into the 'pathoclitic' properties of neurones, although the pathological evidence points clearly to their existence. The position with regard to the 'vascular' factor has been surveyed and it must be admitted that, even now, no adequately verified theory can replace that of Spielmeyer formulated more than 30 years ago.

The conclusion seems unavoidable that the solution of the fundamental problems of hypoxic lesions will not be attained within the discipline of classical descriptive neuropathology but will require the contribution of the physiologist and experimental pathologist who can record such important physiological variables as arterial, venous sinus and intracranial pressures before, during and after an hypoxic episode, the oxygen saturation of arterial and venous blood, the oxygen tension in nervous tissue and the electrical activity of neurones or neuronal systems.

Some of the potentialities of this approach to the aspects of hypoxia have already been demonstrated by the experiments of Meyer and Denny Brown (1955) in the monkey.

The availability of sensitive techniques for the measurement and recording of these intracranial and systemic variables offers to the experimental worker the possibility of escape from the neuropathological deadlock now existing in regard to the pathogenesis of hypoxic lesions.

Acknowledgement. I am most grateful to the Medical Advisory Committee of the National Spastics Society for financial assistance in a current investigation of hypoxic mechanisms.

SUMMARY

Evidence for a period of hypoxia of some type is frequently encountered in the history of cases of cerebral palsy, however variable the final cerebral pathology may be. This hypoxia may arise as a result of, for example, neonatal asphyxia, maternal anaemia, intracranial hærmorrhage or subsequent epileptic seizures.

Both the neurone and the cerebral capillary are damaged by hypoxia but the ultimate cerebral lesions commonly involve a characteristic pattern of selectively vulnerable structures which include cerebral cortex, Ammon's horn, thalamus, amygdaloid nucleus and cerebellum. The various hypotheses advanced to explain this pattern of lesions are discussed with particular attention to that of arterial compression. Reasons are given for believing that it is unlikely that hypoxic lesions can be ascribed to compression of arterial trunks. However, the possibility of arrest of blood-flow at an arteriolar or capillary level deserves consideration.

An understanding of the fundamental mechanism of the hypoxic lesion demands experimental techniques by which physiological variables such as arterial, venous sinus and intracranial pressures and blood and tissue oxygen tensions can be recorded before, during and after a period of hypoxia.

RÉSUMÉ

Dans l'anamnèse des cas d'infirmité motrice cérébrale, on retrouve souvent des traces d'hypoxie d'un certain type, quelque soit la diversité du tableau définitif de la maladie. Cette hypoxie peut résulter par exemple d'asphyxie néonatale, d'anémie maternelle, d'hémorragie intracrânienne ou de crises épileptiques consécutives.

Le neurone aussi bien que les capillaires cérébraux sont lésés par l'hypoxie mais les lésions cérébrales définitives comportent souvent un schéma caractéristique de structures sélectivement vulnérables qui comprennent le cortex cérébral, la corne d'Ammon, le thalamus, le noyau amygdalien et le cervelet. Les diverses hypothèses proposées pour expliquer ce schéma de lésions sont discutées et tout particulièrement celui de la compression artérielle. L'auteur donne des raisons de croire qu'il est improbable que les lésions hypoxiques puissent être attribuées à la compression des troncs artériels. Cependant, la possibilité d'un arrêt du flux sanguin au niveau artériolaire ou capillaire mérite d'être envisagé.

Il semblerait que, pour comprendre le mécanisme fondamental de la lésion hypoxique, il faille des techniques expérimentales permettant d'enregistrer les variables physiologiques telles que les pressions artérielles, des sinus veineux et intracrâniennes, ainsi que la tension en oxygène du sang et des tissus, avant, pendant et après la période d'hypoxie.

ZUSAMMENFASSUNG

Man findet oft in der Anamnese der Fälle von Zerebrallähmung Spuren von irgend einer Art von Hypoxie, wie verschiedenartig das endgültige Krankheitsbild auch sei. Dieser Mangel an Sauerstoff kann zum Beispiel als Folge neonataler Asphyxie, mütterlicher Blutarmut, Hämorrhagie innerhalb der Schädelhöhle oder späterer epileptischer Anfälle erscheinen.

Sowie die Neurone als auch die Gehirnkapillaren sind durch Hypoxie geschädigt aber die endgültigen Läsionen des Gehirns weisen oft eine charakteristische Schablone von besonders verwundbaren Strukturen auf, die die Hirnrinde, das Ammonshorn, den Thalamus, den amygdaloiden Nucleus und das Kleinhirn einschliessen. Die verschiedenen Hypothesen die vorgeschlagen wurden, um diese Läsionsschablone zu erklären, werden erörtert, mit besonderer Rücksicht auf die Arterienkompression. Der Autor erklärt, warum man annehmen kann, dass es unwahrscheinlich ist, dass Schädigungen durch

Hypoxie einer Kompression der Arterienstämme zugeschrieben werden können. Man muss jedoch die Möglichkeit einer Stockung des Blutflusses im Gebiet der kleinen Arterien oder Kapillaren in Betracht ziehen.

Um den Grundmechanismus der Schädigung durch Hypoxie zu verstehen, wären, dem Anschein nach, experimentale Techniken nötig, um physiologische Befunde wie Arterien -Sinus- und Gehirndruck, sowie Sauerstoffgehalt des Blutes und der Gewebe vor, während und nach der Periode der Hypoxie, zu registrieren.

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Lower Extremity Bracing In Cerebral Palsy*

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TREATING cerebral palsied children offers far more challenge to the physician today than a decade or two ago. The attention and study directed to cerebral palsy in the United States and elsewhere have resulted in broader treatment concepts and indeed a more basic approach in many phases of this disease. Fresh ideas, so often unsettling to established routines, deserve a reception both welcoming and critical. Here are new criteria against which to measure our concepts, new procedures to cause us to scrutinise our own. In this light let us re-examine one of our established methods —bracing.

The transitional stage in which we find cerebral palsy treatment today may be traced in origin to the experimental work of Sherrington (1947) and his pupils. Within recent years Temple Fay (1948) and Herman Kabat (1947), pursuing somewhat different leads, have sought clinical application for the increasing knowledge of reflex mechanisms. These men visualised the central nervous system as a complex repository of reflex and patterned movements capable of release into functional channels. This was to be accomplished through carefully evolved treatment techniques based on prolonged patient training. Without dwelling on the historical process or attempting to credit the many individual workers, we may briefly consider a few of the principles underlying the neurophysiological approach to treatment. Parenthetical references are supplied to indicate how a principle has been applied; they are not meant to characterise an author's concepts.

* Read in Spanish at the Sixth National Congress of Orthopedics and Traumatology, Mexico City, April, 1960.

Inhibition

Before functional training of trunk and extremities can begin it is often necessary, at least with severely handicapped children, to establish control of interfering postures and movements. This is the basis for the relaxation concept underlying the classical treatment of athetosis. Interfering movements may be of bewildering variety, and efforts to eliminate—or rather limit—them have produced a wealth of technical approaches. So far as these methods can be summarised, reflex control is sought through *position* (control of otolith and neck afferents: Phelps' technique; *see* Egel 1948), *posture* (reduction of tonic inflows: *see* Fay 1954, Bobath and Bobath 1956), *controlled movement* (utilisation of segmental inflows; *see* Kabat 1950), or *skin stimulation* (utilisation of dermatome afferents; *see* Rood 1956).

Facilitation

Like inhibition, a function with which it is closely identified, facilitation is met with at all levels of central nervous system function: cortical, cerebellar, brain-stem, spinal. In the treatment of cerebral palsy facilitation may be elicited at higher (Fay 1954) or lower (Kabat 1950) levels of the neuraxis; or through proprioceptive mechanisms (Bobath and Bobath 1956).

Integration of Associated Movements

Many of our common movements are subserved by nervous system patterns which are never consciously learned. We observe in the infant, for instance, gradual improvement in the ability of the head to follow the direction of gaze, and of shoulder movements to accompany hand placement. Associated movements in patients with

cerebral palsy are often aberrant, the effort of the therapist being to redirect them into functionally useful patterns (Semans 1959).

Attainment of Balance

Control of equilibrium for sitting, standing and walking is a basic objective of cerebral palsy treatment. There is little we can expect of a child whose world gyrates about him. For severely quadriplegic children balance is always hard-won, sometimes even unattainable. Poorly oriented in space because of deficient labyrinthine and proprioceptive inflows, their postural tonus and righting reactions impaired, these patients must additionally contend with phasic movements and tonic reflexes which constantly threaten whatever precarious equilibrium they manage to achieve. It becomes our concern to ensure stability if we expect to get very far with other aspects of treatment.

Principles of Treatment

With this brief sketch of neurophysiological considerations the rationale of lower extremity bracing in cerebral palsy may in part be indicated. A few of the principles directing our general approach to treatment of children with this disease should first be stressed:

1. Treatment centres in the child's home, the effort being to provide a social and educational environment approximating to that of a normal child.
2. The goal of treatment is to develop useful function as promptly as possible, using methods which make patient and parents active partners in the rehabilitation process.
3. Incentive being the therapist's greatest ally, any motor effort not clearly harmful to the child deserves encouragement. Present knowledge scarcely justifies doctrinaire approaches, such as insistence that motor training must recapitulate phylogeny.
4. Posture precedes movement; balance precedes walking.
5. Multiple aberrant movements are to be reduced and simplified for training purposes.
6. Training emphasises patterns of movement, assimilating these into functional and recreational activities.

Braces can be made to serve a very useful role in any treatment of cerebral palsy based on concepts such as these. With large numbers of patients various types of braces are needed for different purposes. One model which in our

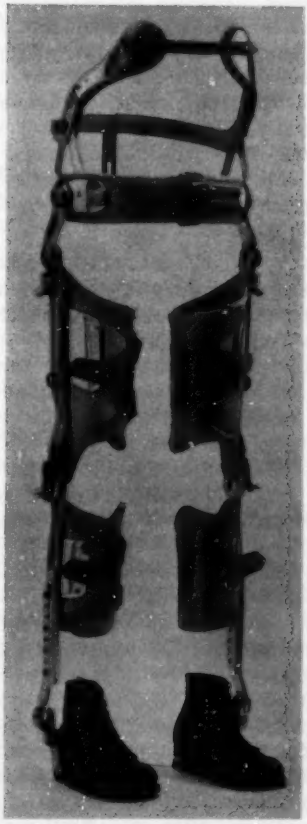

Fig. 1. The Newington Brace for Cerebral Palsy is made largely of aluminium alloy. Light in weight, its design assures support to the patient while incorporating maximal resistance to the stresses of involuntary movement.

experience has proved durable, effective and adaptable is the Newington Brace for Cerebral Palsy (Fuldner and Rosenberger 1958). The rationale for using a brace of this type (Fig. 1) may be stated as follows:

A well-made brace provides an effective mechanism for *inhibition*. This applies not only to restraint of obvious aberrant movements, but also to control of stretch reflexes generally. Uncontrolled stretch (and flexor) responses underlie a number of handicaps in cerebral palsy—toe thrust, for example. One often sees attempts to overcome a stretch reflex by repetitive manual stretching. No treatment could be more illogical in concept or more apt to aggravate what it purports to relieve. While repetitive stretch increases muscle irritability, sustained stretch 'breaks through' the myotatic reaction and seems eventually to raise the threshold for its appearance, perhaps by producing elongation of the tendon.

Braces may also be used for *facilitatory* effects. A familiar example is the immediate improvement in the hand co-ordination of athetoid children whose legs and trunk are stabilised by bracing. A brace constructed with multiple locks facilitates discrete joint control. Reciprocal hip movements, for instance, are more readily taught when the distracting effects of interfering reflexes from other parts of the body can be blocked off. For training sessions this involves trunk support (pelvic band) and knee stabilisation (knee locks). The patient thus becomes relatively free to direct attention to the immediate training objective.

This technique also illustrates the general approach to *control of associated movements*. Thus, dissipation of synergies concerned in reciprocation patterns can be offset by locking out overflows until the patient learns how to co-ordinate proximal leg movement for whatever type of gait is being taught. After the central pattern is understood and reasonably stabilised, movements peripheral to the pattern can be gradually 'fed back', emphasis now shifting to their control and utilisation. The skilful therapist directs training along functional lines to maintain interest—hers and the child's. In cerebral palsy, as elsewhere, a monotonous routine is usually an unproductive one.

Lower extremity bracing finds one of its most useful applications in *balance training*. The balance-deficient child is handicapped in all aspects of treatment: not solely in trunk carriage and gait, but in upper extremity control as well. Imbalance in cerebral palsy is frequently of complex origin. Righting reactions and check movements may be absent, proprioception impaired, vestibular connections damaged. Experimentally, it is known that posture is in large part mediated reflexly, by way of brain-stem mechanisms; righting reactions, for example, are not appreciably affected by removal of the cerebral hemispheres (Evans 1956). This is confirmed by what one observes of balance development in normal as well as brain-damaged children. Balance is acquired through gradual integration of reflex mechanisms, a process in which consciously willed movement appears to play little part. In this acquisition the significant difference between the cerebral palsied child and his normal counterpart lies in the defective nervous system pathways which the former must utilise.

Balance training therefore emphasises placement and support of the patient in such a way as to take advantage of reflex mechanisms available and assist in their progressive integration. An adaptable brace permits us to focus training initially on the head and neck. As control is achieved here, the brace allows emphasis to shift successively to trunk, pelvis and lower extremities. The child not only practices balance but is introduced to the concept of free stance and the possibility of walking as he sees other children walking. Fear of collapse is removed. Braces may also function by substituting dermatome afferent impulses for deficient proprioception, as in body righting reflexes demonstrable in labyrinthectomised animals (Fulton 1955; Fig. 2).

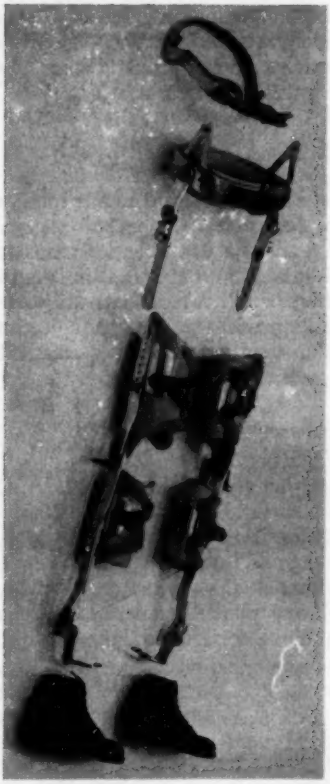

Fig. 2. The brace is readily disassembled. This gives the physician considerable latitude in adapting it to the child's varying needs in training and support. Sections may be removed during training periods, and discarded as voluntary regional control is attained.

Other Uses of Braces

This concludes our brief survey of neurophysiological aspects of bracing. Reference should be made in closing to the many further ways in which braces can be helpful in cerebral palsy. They are the most effective means of preventing contractures—of tendo achillis and hamstrings, for example—and of maintaining axial alignment of the lower extremities. From this aspect braces may be used prophylactically for the paraplegic child who habitually assumes deforming postures, such as the common one of sitting on the knees with legs spread to the side. At the proper time bracing will get on their feet those balance-defective children who insist on crawling as a means of locomotion. Braces are usually preferable to casts for long-term maintenance of alignment following operative procedures. An important asset of bracing as a form of therapy is its constant availability, enabling us to extend the usefulness of time-limited treatment sessions. Night bracing, for example, may be very helpful in some instances. When we include bracing in our rehabilitation programme two potential therapists become available for 24-hour duty—the child's parents.

Conclusion

It is scarcely necessary to remind the readers of the *Bulletin* that bracing is an adjunct, not a system, of treatment. Whatever programme we follow in treating a cerebral palsied child let us not fall into the common fallacy of ascribing the improvement to the system. If we never treated the child at all he would, in some respects, improve. In others he might very likely get worse. The essence of treatment, in our view, is to assist and direct the immense inherent potential for improvement, at the same time striving to prevent and correct the untoward maturation concomitants of a faulty nervous system. In this approach braces can be of inestimable benefit. For the physician possessed of patience, imagination and reasonable skill they serve as versatile instruments of wide adaptability in the treatment of cerebral palsy.

SUMMARY

Established methods of treating cerebral palsy are re-examined in the light of recently introduced techniques based on neurophysiological approaches. These aim to control involuntary movements and encourage voluntary activities through the utilisation of reflex

mechanisms in the central nervous system. Inhibition, facilitation, integration of synergic movements and balance attainment are among the principles employed. The same principles can be constructively applied to bracing as a therapeutic technique. Properly made, a lower extremity brace is a highly effective adjunct in the treatment of cerebral palsy.

RÉSUMÉ

L'appareillage d'un membre inférieur dans l'infirmité motrice cérébrale.

Les méthodes de traitement de l'infirmité motrice cérébrale sont reprises à la lumière des techniques récentes procédant d'approches neurophysiologiques. Ces techniques, par les mécanismes de reflexe du système nerveux central, tendent à contrôler les mouvements involontaires et à stimuler les activités volontaires. L'inhibition, l'aisance, l'intégration de mouvements synergiques et l'acquisition de l'équilibre sont, notamment, utilisées. Ces mêmes principes peuvent être utilement appliqués à l'appareillage en tant que technique thérapeutique. Correctement exécuté, l'appareillage d'un membre inférieur est un auxilliaire de grande efficacité dans le traitement de l'infirmité motrice cérébrale.

ZUSAMMENFASSUNG

Korrektierapparate der unteren Extremitäten bei Zerebrallähmung

Die Behandlungsmethoden der Zerebrallähmung werden vom Gesichtspunkt der neuen, auf physiologische Basis gestützten Techniken aus, untersucht. Diese Techniken erzielen die Beherrschung der unwilligen Tätigkeit durch die Reflexmechanismen des Zentralnervensystems. Hauptsächlich werden Inhibition, Erleichterung und Integrierung der synergischen Bewegungen und Erwerbung des Gleichgewichts verwendet. Dieselben Prinzipien können erfolgreich für die Apparate als Behandlungstechnik angewendet werden. Ein geeigneter Apparat ist eine höchst wirksame Hilfe für die Behandlung der Zerebrallähmung.

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The Child's Hearing For Spoken Language

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THE terminology of disorders of auditory communication and impaired hearing in young children is confused. In a previous paper (Sheridan 1959) I offered my own definitions, as follows:

Language is the symbolisation of thought in speech, gesture or writing, for the purpose of interpersonal communication. (Thus the term language also includes communication through reading and writing which are outside the theme of this paper.)

Speech is the use of systemised vocalisations to express verbal symbols or words.

Hearing is the reception of sound by the ear and its transmission to the primary auditory area of the cortex.

Listening is the act of paying attention to the sounds received, with the object of interpreting their meaning.

Interpretation of spoken language depends on previous *learning*—that is, on the gradual building up of an efficient memory register, to which incoming words can be referred for matching (i.e., *recognition*), and from which needed words can be summoned at will (i.e., *recall*).

There also exists between young infants and their mothers a pre-linguistic, non-symbolic interpersonal communication which expresses itself on the child's part in clinging or nestling, in resistive stiffening or vigorous bodily activity, and in unorganised but emotionally-charged vocalisations. It is concerned with the infant's immediate basic needs and feelings. The mother soon becomes adept at interpreting it. Similarly the older non-speaking but ambulant child makes known his needs by pulling his mother in the direction he wishes her to go, placing her hand on an object he desires, and pointing urgently with his finger; or he expresses his feelings by stroking her face caressingly, smiling, screaming or throwing a tantrum.

This primitive form of communication deserves closer study than it has yet received because, when the 'shorthand' of linguistic or mimed symbolism fails him, the child must resort to it in order to maintain contact with his fellow-beings. It is necessarily concerned only with the here and now. It cannot express past or future tense. Nevertheless, it provides a basis of comprehensible communication between the child and his familiars, and might possibly be extended and systematised so that the non-mobile, non-speaking child, who must sometimes be left for days or weeks in the care of other people, need not be left entirely without means of communication.

Spoken Language

Spoken language implies communication between two brains. Reduced to simplest terms, the functions of the brain with relation to spoken language may be regarded as three-fold; reception, which begins in a sensory stimulus; interpretation, which is an intellectual process with emotional implications (or involvements); and expression, which terminates in motor activity. The neurosurgeons have been able to outline with electrodes certain areas of the brain which are intimately concerned with hearing, remembering and producing spoken words (Penfield and Roberts 1959). It has not been possible to discover how the young child can combine his diverse powers of perception, symbolism and impulse to motor activity so as to produce spoken or mimed language (Critchley 1960). Yet the instinct to communicate in speech and gesture is

innate. Reading and writing, on the other hand, must be voluntarily learnt.

Reception depends primarily on hearing. The ear can never 'switch off', nor has it any power of selection. It is obliged to relay to the temporal lobe whatever medley of sounds is received by the cochlea. From then onwards the mind takes charge. The first step is decision to listen to what is being said. This depends not only on willingness but also on capacity to do so. In cerebral palsy, for instance, the wish may be present, but distractibility is excessive so that the child is unable to pay adequate attention. The willing and capable brain then rapidly proceeds to select the significant or 'foreground' speech elements from the general background of noise, sorts them out in appropriate tonal and temporal patterns, refers all this mass of raw material to the memory register for matching against previously recorded patterns, and finally, in the light of intellect and the warmth of emotion, decodes the message. Thus *interpretation* depends on many stored memories, not only of the verbal symbols themselves and of the people, objects, activities and trains of thought they represent, but also of countless related emotional experiences. If reply is needed, *expression* is inaugurated by another act of mental decision (again involving will and capacity), after which comes the marshalling of relevant ideas, the summoning up from the memory register of verbal symbols suitably invested with grammatical form and appropriate vocal cadence, the assembly of kinaesthetic images, and finally the motor impulse which puts the required muscles in action. The process is not yet over, however, because, as the listener receives his message, the speaker simultaneously receives a feedback into his own central nervous system of a host of stimuli—auditory, vibratory, kinaesthetic, visual, emotional and thought-provoking.

Normal Development of Hearing and Speech

The normal development of meaningful hearing in the child depends on a number of biological and environmental factors. *First*, the child must possess normal anatomical structures for receiving sound, and the cellular components of these peripheral and cerebral structures must be continually supplied with the biochemical agents necessary for normal development and function. *Second*, the child must possess the power to attend to and to decode the sound signals received. This ability depends partly on the child's opportunity to build up a store of auditory memories so that he has a sufficient number of patterns with which to match any incoming sound against his previous experiences of it, and partly on his native intelligence, which enables him to make the appropriate reference quickly and judiciously. In other words, his ability to interpret depends upon inborn capacity and environmental opportunity. *Third*, the child must have his natural incentive to establish auditory rapport with his fellows continually reinforced in the warmth of favourable emotional 'climates'. Thus, hearing with comprehension, like every other function of the mind, depends on *maturation* and *learning*.

At birth the cochlea of the normal baby has already reached an advanced stage of anatomical development. In fact, it is one of the most fully developed of all the body structures. Highly efficient apparatus for the reception of sound is therefore present, but the interpretative cortex is still neurologically immature and has everything to learn.

The neonate's responses to sound are entirely reflex. During this early period the child is by no means silent. He cries loudly when he is uncomfortable and utters little gutteral noises when he is content. These vocalisations are also reflex and they occur in deaf as well as hearing babies. Within a few weeks he shows signs of his growing awareness of nearby meaningful sounds. Literally he learns to listen and to speak on his mother's knee. He rapidly comes to associate the touch of her hands and the sight of her face and the cadences of her voice with the comfort she brings him. At

about 3 to 4 months, his immediate response to the appearance of a nearby face is to smile, activate his limbs and vocalise with pleasure. As Rheingold and others have shown (1959), these vocalisations are reinforced or extinguished, according to the vocal response of the adult.

By 7 to 8 months, the normal child's ability to attend to and localise the meaningful homely sounds that occur within his spatial world is easily demonstrated. He also babbles, laughs and squeals loudly and purposefully. During the same period, the deaf child's vocalisations gradually diminish and by 7 to 8 months have become few and toneless, although he may be so visually alert and socially attractive that his meagre vocalisations and his lack of response to sound may be overlooked.

About 9 to 10 months, the normal child's response to the speech of his familiars is very obvious, and he babbles incessantly, using a wide range of vocal cadences. At 10–12 months he shows, by his behaviour, that he understands much of the simple language addressed to him, and he can mimic his mother's playful vocalisations, including some word-forms (i.e., imitation). A month or two after his first birthday, he begins spontaneously to use recognisable words, usually first by naming familiar objects (i.e., recognition). By about 18 months, he can call to mind the verbal symbol in the absence of the object (i.e., recall). This marks the beginning of true language. At 2 years the normal child is putting words together to form little sentences. His mother, listening with the ear of love and familiarity, will assert that 'he can say anything', but, since he uses a number of infantile mispronunciations, his speech is not easy for outsiders to follow. Nevertheless his linguistic development is usually sufficient to allow experienced testers to apply various vocabulary tests with toys and pictures. At 3 years, he has a large working vocabulary and most of what he says is intelligible even to strangers. From this age, it is possible for the experienced tester not only to apply a standardised clinical test but also to obtain a useful pure-tone audiogram. It needs to be emphasised that the procedures employed in these tests, however simple they may appear, demand high skill and will only produce valid results when they are suitably applied by people who understand the acoustic and psychological principles underlying them.

In the natural development of spoken language, therefore, there is a comparatively long time-lag between learning to listen and learning to talk. During the first year of life the child must store his auditory memories from stimuli applied at mother-distance—i.e., close to his ear—at high decibel level, in words which are addressed to him personally, and which concern events happening to him in the immediate present. At the same time he must be given strong encouragement to vocalise in return.

It is often said that the young child speaks as he hears. It would perhaps be more accurate to say that, because his performance lags behind his perception, his spoken language echoes what he appreciated through his hearing some months previously. This is an important point to stress in auditory training and speech therapy. The child will always be able to recognise and repeat a speech sound accurately for some time before he can call it up from his memory and use it correctly in spontaneous speech.

If for any reason, such as impaired hearing, mental backwardness, separation or rejection, he does not receive enough of this intensive individual coaching in his mother-tongue to fix it securely in his memory during the period which is critical for learning it—i.e., in the first 3 years, and most particularly the first 1½ years—his difficulties in acquiring spoken language will be seriously increased. This is partly because once the child is mobile his exploratory interests will absorb more of his attention, and his mother's voice will be heard from greater distance and probably less often addressed directly

to him. Moreover, the emotional bonds between the infant and his mother and the relationship between the toddler and his mother are totally different. She uses new and different speech forms and tones to the older child. The emotional qualities of speech depend on countless delicate variations of vocal tune and stress and can only be adequately appreciated if they have been heard frequently in context. Before the advent of electronic hearing-aids, deaf children can have had little conception of the significance of emotional overtones.

The Hearing Needed for Speech

Speech sounds, in common with other varieties of sound, possess attributes of pitch, intensity and duration. The human ear is capable of hearing about 10 octaves in pitch. The speech range covers some 7½ octaves, from about 100 to 10,000 cycles per second. Broadly considered, the frequencies below 1,200 cycles provide most of the carrying power, energy and emotional colour of speech (Fry 1957) while those above 1,200 cycles are concerned with intelligibility. The chief characteristic components of all the vowel sounds, except the long *ee* and its shorter form *i*, which have significant components in the upper frequency bands, are in the lower and middle frequencies. Some idea of how the vowel scale rises in pitch from *oo*, the lowest vowel sound, to *ee*, the highest vowel sound, may be obtained by speaking aloud or recalling mentally the sounds *oo*, *oh*, *aw*, *ah*, *eye*, *ay* and *ee*. The chief components of all the consonant sounds are in the middle and high frequencies. The principal components of the nasal sounds, *m*, *n* and *ng*, border on the low frequencies. Those of the *r*, *l*, *w*, *y* group of continuants are higher in the middle frequencies. The plosives, *g*, *d*, *b*, and their voiceless equivalents, *k*, *t*, *p*, are still higher in scale, bordering on the upper frequencies. The sibilants, *ch*, *sh*, *zh*, *z*, *s*, and the fricatives, *v*, *f*, *th*, are the highest of all in pitch. The fundamental vowel pitch of a woman's voice is about an octave higher than a man's (i.e., in the region of 256 cycles). Children's vowels are still higher in pitch. The highest consonant components of both male and female voices, however, are practically the same.

The decibel level at which a vocal sound is delivered determines its loudness and carrying power. The vowels possess higher decibel intensity than the consonants, the sound *aw* possessing the strongest and the sound *ee* the least of the vowel intensities. The sibilants and fricatives possess the lowest decibel intensity, and therefore have a weak carrying capacity. The weakest sound of all is the voiceless *th*. The difference in intensity level between *aw*, the strongest, and *th*, the weakest phonetic unit is approximately 30 decibels. The sibilants and fricatives are therefore highest in pitch and weakest in intensity. This causes them to be difficult to distinguish if conditions are in any way unfavourable.

Thus the interpretation of the simplest spoken phrase imposes on the listening brain the necessity to attend instant by instant to a large number of complex sounds which swing rapidly over differences of some 8 octaves in pitch and 30 decibels in intensity, and finally to synthesise the sounds heard into a meaningful whole. For practical purposes one may assume that a quiet conversational voice at 3 ft. carries to the listening ear sound-intensities varying between peaks at 60 db and troughs at 30 db.

The tunes of speech, the stressing of syllables, the timing of vowel sounds, and the crispness of consonant articulation all add to the liveliness and intelligibility of speech.

The nature of stereophonic hearing is still very imperfectly understood, but the fact that vocal communication normally occurs in three-dimensional space must have a bearing on the child's acquisition of spoken language.

The Deaf Child

The child with impaired hearing over any part of the speech range will receive, and therefore memorise and recall, imperfect phonetic patterns. His own speech will

be correspondingly distorted. In high-tone deafness the consonant frequencies are mainly affected. The child may therefore begin to vocalise and use vowel sounds at the usual age, so that his hearing disability may remain unsuspected although his speech continues to be unintelligible. These children run a real risk of being considered mentally defective (Minski 1957, Sheridan 1944). Severe loss over the middle and lower frequencies will prevent the spontaneous acquisition of spoken language altogether. Provided the interpretative faculties are intact, the young child can make excellent use of a limited field of hearing, but only if he is given individual expert auditory training at close quarters with an efficient hearing-aid in the critical period (Whetnall 1956, 1958, Ling 1959). The need to compensate for the period of auditory deprivation before training began must always be explained to the parents. Otherwise they will be disappointed and perhaps fail to persevere because the child does not begin to speak immediately he is fitted with an aid.

Need for Tests of Vision and Intelligence

The full assessment of the child's capacity for spoken language does not end with the application of clinical and audiometric tests, otological examination and the provision of a hearing-aid. In the normal acquisition of speech not only the child's hearing but his vision and his intelligence are closely implicated.

The testing of vision in normal preschool children is comparatively easy when suitable material is used and the examiner is experienced (Sheridan 1960). The cooperation of deaf children who have normal intelligence is readily assured. Considerable difficulties are encountered, however, when the child is not only deaf but mentally retarded, highly distractible, emotionally maladjusted, or subject to uncontrollable movements.

Further difficulty arises in the application of intelligence scales. The estimation of intelligence in deaf children, and particularly deaf children with an additional handicap, is never easy, and often exceedingly difficult. All one can truthfully say in these cases is that the child's intelligence quotient is not less than a certain figure, with the implication that it may be appreciably higher. The diagnosis of ineducability in such children can never safely be made on the results of one interview, or by inexperienced examiners. It must be kept in mind that dysphasia of any sort may be accompanied by dyslexia and dysgraphia. This is particularly important in cerebral palsy.

Auditory Training in Cerebral Palsy

The instinct to communicate in spoken language is very strong, and, as has been said, when the interpretive faculties are intact, the majority of young children with cerebral palsy and impaired hearing may be expected to make good use of auditory training. Nevertheless, there are a few deaf children with and without cerebral palsy who demonstrate a promising-looking audiogram, average performance on non-verbal intelligence tests, and little or no motor disability but cannot acquire useful speech. In some cases they cannot learn to lip-read. It is as if they lack the ability to perceive meaning in spoken words, either through their hearing or through their vision. It is possible that this indicates some disturbance of the brain's timing mechanisms, producing an auditory-temporal disorder analagous to the well-known visual-spatial disorders.

Impairment of auditory perception, particularly for high-pitched sounds, is common in athetoid cerebral palsy. It is often impossible to decide whether the deafness and gross disorders of spoken language associated with athetosis are the result of damage to the auditory, motor or interpretive functions of the brain, or to a combination of all these. When a significant hearing loss is demonstrable or surmised, otological opinion should immediately be sought and auditory training instituted by an experienced teacher of the deaf.

Speech Therapy

The whole question of the treatment of defective spoken language in cerebral palsy needs review. Much time and energy are now devoted to empirical procedures that are almost exclusively concerned with motor aspects of respiration, deglutition and articulation. Conscientious study of the available literature and discussions with many speech therapists working in this field have failed to elucidate what guiding principles are followed in treating the child's disorder of spoken language. There is little agreement on practice, and still less on theory. Nor is it easy to assess results. Some children do not improve after years of treatment, while others improve without any. The therapist's time might be more profitably spent in devising improved methods of communication for each individual child, and in giving appropriate instruction to his parents and teachers. At present the speech therapist receives little help from doctors, psychologists or teachers. A concerted scheme of research under the leadership of a paediatric neurologist is urgently needed. It is vitally important, for instance, to discover whether and to what extent spoken language that is adequate for everyday purposes can be produced by training after 7 years of age if it has not already appeared spontaneously before that time; and at what stage concentration on routine speech therapy should be abandoned in favour of teaching some alternative means of communication.

Conclusions

The ideal of enabling every deaf child (with or without an additional handicap) to use spoken language as a matter of course must always be the first aim of treatment and training. But if, after adequate trial, this proves unattainable, every effort must be made to provide the child with some means of communication which will be effective with the largest number of persons and in the largest number of everyday situations.

On the ability to communicate with one's fellows depends not only one's learning and livelihood but one's happiness as a human being.

SUMMARY

An attempt is here made to clarify the confused terminology of disorders of communication and to differentiate between speech and language and between hearing and listening.

The interpretation of spoken language depends on the adequate storage of auditory memories in the central nervous system, and on the subsequent ability of the mind to recognise and recall verbal symbols from the memory register. Thus the development of spoken language depends on maturation and learning.

Failure to acquire spoken language may arise from (1) lack of opportunity to learn in the early years, which are critical for its development; (2) impaired hearing; (3) general intellectual inferiority; (4) damage to or failure of development of the parts of the brain concerned with the interpretation and manipulation of language symbols; and (5) lesions of the peripheral speech mechanisms.

The physical properties of speech sounds are briefly considered, with special reference to the parts of the auditory range that are concerned with the hearing of vowels and consonants.

If the outstanding difficulties of speech therapy in relation to defects of spoken language in cerebral palsy are to be solved, there is a need for a new team approach, under the leadership of a paediatric neurologist.

RÉSUMÉ

Comment un enfant entend le langage parlé.

L'auteur tente de clarifier la terminologie confuse appliquée aux troubles des moyens de communication et de faire une distinction entre la parole et le langage et entendre et écouter.

L'interprétation donnée au langage parlé dépend de la réserve adéquate en souvenirs auditifs dans le système nerveux central et de la faculté consécutive qu'a l'esprit de reconnaître et de rappeler les symboles verbaux du registre de sa mémoire. Ainsi le développement du langage parlé tient à la maturation et à la faculté d'entendre.

L'insuccès dans l'acquisition du langage parlé peut être causé par: (*1*) l'absence d'occasion d'apprendre au cours des premières années si décisives au développement de l'enfant; (*2*) une audition défectueuse; (*3*) une pauvreté générale de l'intellect; (*4*) un développement amoindri ou nul des secteurs du cerveau gouvernant l'interprétation et l'emploi des symboles du langage; (*5*) des lésions des mécanismes périphériques de la parole.

Les propriétés physiques des sons émis par la parole sont brièvement considérées avec mention particulière pour les secteurs du champ auditif intéressés par l'audition des voyelles et des consonnes.

Si les difficultés dominantes de la thérapie du langage relative aux défauts du langage parlé dans l'infirmité motrice cérébrale doivent être résolues, il faut reprendre le problème au moyen d'un travail en équipe sous la direction d'un pédiatre neurologue.

ZUSAMMENFASSUNG

Wie ein Kind die gesprochene Sprache hört.

Der Autor versucht, die verwirrte Terminologie der Störungen der Verbindungsmittel zu klären und zwischen Rede und Sprache, hören und horchen zu unterscheiden.

Die Deutung der gesprochenen Sprache hängt von dem angemessenen Vorrat von Gehörerinnerungen im Zentralnervensystem und von der folgenden Fähigkeit des Geistes, die Sprachsymbole zu erkennen und aus dem Gedächtnissregister zurückzurufen, ab. Die Entwicklung der gesprochenen Sprache hängt also von der Reifung und der Hörfähigkeit ab.

Ein Misserfolg in der Erwerbung der gesprochenen Sprache kann von folgenden Umständen verursacht sein: (*1*) Mangel an Gelegenheit zu lernen währen der ersten Jahre, die so entscheidend für die Entwicklung des Kindes sind; (*2*) ein schlechtes Gehör; (*3*) ein allgemeiner Intelligenzdefekt; (*4*) Beschädigung oder Ausbleiben der Entwicklung der Gehirnbezirke, die an der Auslegung und der Verwendung der Sprachsymbole beteiligt sind; (*5*) Läsionen der peripherischen Mechanismen der Rede.

Die physikalischen Eigenschaften der Sprechlaute werden kurz betrachtet, mit besonderem Hinweis auf die Bezirke des Hörfeldes, die am Hören der Vokale und der Konsonanten beteilig sind.

Wenn man die hauptsächlichen Schwierigkeiten der Sprachtherapie in Verbindung mit Defekten der gesprochenen Sprache bei Zerebrallähmung beseitigen will, ist eine neue kollektive Arbeit unter der Leitung eines Kinderneurologen nötig.

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Hearing and Speech Disorders in Childhood*

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Success in the training and education of deaf children largely depends on finding them young. This can be greatly facilitated by recognising which babies are 'at risk' because they or their mothers have been exposed to known causes of deafness. By analysing these causes it is possible to draw up a register of all babies in a community who are 'at risk'. This should be done in all big hospitals, probably by the paediatrician.

THE early diagnosis of a hearing loss in children and the management and education of those suffering from this handicap are arousing increasing interest, not only among the medical and education authorities but also among the general public. With the improvement in hearing-aids and amplifying equipment more can now be done to alleviate this appalling handicap, and there is hope that with training and education these children will take their place in the world as useful citizens making full use of their capabilities.

I am not dealing here with conduction deafness brought about by upper respiratory infections (though this may cause great inconvenience in schooling because it may vary from day to day) but only with the problems of children who are born deaf or lose their hearing very soon after birth. Unless these children are given special attention and training they cannot acquire speech and all the benefits that speech and language afford.

A child with normal hearing imitates sounds and learns to interpret them during the first three years of his life. Unless a child with a severe hearing loss has his residual hearing trained during these early years he will have the greatest difficulty in learning words and language later. Most so-called deaf babies have some residual hearing, and if this can be trained with the help of hearing-aids and other amplification during those early critical years the child will have his best chance of learning speech and language.

*Read to the Section of Paediatrics, Royal Society of Medicine, on February 26, 1960.

Ascertainment

The first problem is how to find these handicapped children when they are very young. A study of the causes of their deafness will show that certain babies must be considered 'at risk' or 'suspect'.

CAUSES OF PERCEPTION DEAFNESS IN INFANTS

PRE-NATAL
- Heredity
- Diseases of pregnancy: virus infection, toxaemia
- Abortifacients
- Prematurity
- Hazards of labour: anoxia, birth trauma
- Congenital anomalies: atresia (middle-ear deafness)

NEO-NATAL
- Anoxia, prematurity, post-maturity (?) and birth-injury
- Kernicterus of Rhesus incompatibility
- Kernicterus of prematurity

POST-NATAL
- Acute infections: measles, mumps, meningococcal / pneumococcal / influenzal / tuberculous } meningitis
- Toxic drugs: streptomycin

UNKNOWN
- Accounts for 30 per cent of all deaf babies

In the pre-natal group, the virus infection contracted by the mother in pregnancy need not be rubella; influenza and measles are both important. Of the neonatal causes, the dangers of blood-incompatibility are fortunately becoming steadily less. Similarly in the post-natal group, streptomycin may have accounted for some cases but this was more true in the past than now.

The 'unknown' group accounts for about 30 per cent of all deaf babies.

Two recent studies of the hearing of children give some interesting figures for 'at risk' babies. Dr. B. Humphries (1954), a medical officer of health at Leicester, arranged for the health visitors to test the hearing of 4,000 children under 3 years of age. Out of these 4,000, only 4 children were found to have any hearing loss, and 3 of the four were already 'suspect' because their parents were deaf. In the second paper, Dr. Irene Howarth (1958), chief assistant medical officer of health for Lancashire, gives the findings of hearing tests done on 662 'suspect' or 'at risk' children, of whom 6 were found to have a perceptive deafness. She also tested a control group of 3,000 non-suspect children and found that only 2 of them had a similar hearing loss. Thus, according to her figures, there is a 14 times greater chance of finding a hearing loss among suspect children than among non-suspect ones. This is enough evidence to justify the repeated testing of all 'at risk' babies until the presence or absence of a hearing loss is confirmed.

For this purpose a register of 'at risk' babies should be kept at all large hospitals and made as complete as possible. Most of the 'at risk' babies will come, at their first attendance, to the paediatric department, so the paediatricians would seem to be the best people to keep the register, if they are not already doing so, and to follow up the cases at their ordinary outpatients. Information should come in from the medical officers of health, who should be kept informed through their maternity and child-welfare services and health visitors. General practitioners should be encouraged to report to their M.O.H. any cases of virus infections contracted in early pregnancy.

Many children will still slip through the mesh and their hearing loss remain for a long time unrecognised, unless more publicity is given to the probability of deafness in these cases. Both doctors and the lay public must also be made aware that backwardness and late development of speech may be due to a hearing defect. The mother is often an unreliable witness, but, to quote Ballantyne (1958), 'The mother who thinks her child is deaf is rarely wrong.' If a child is suspected of being deaf, its hearing must be investigated repeatedly until it is certain that there is no hearing loss.

For the three years, 1957, 1958 and 1959, the number of new cases of perception deafness in children under 5 years of age seen at Oxford, Reading and High Wycombe, three towns within a 30-mile radius of one another, were respectively 21, 29 and 12, totalling 62. (I am indebted to Mr. Hunt Williams for the Reading figures.)

In 1960, the number of cases seen at the Radcliffe Infirmary, Oxford, was 118, made up of 58 children under 5 years of age, 50 between 5 and 15 years, and 10 cases of bilateral congenital atresia in children under 10 years. The causes of deafness in these 118 cases can be analysed as follows:

CAUSES OF 118 CASES OF PERCEPTION DEAFNESS IN CHILDREN UNDER 15, SEEN AT RADCLIFFE INFIRMARY, 1960

	Cases	%
PRE-NATAL		
Heredity	16	14
Diseases of pregnancy: virus infection and toxaemia	21	17
Prematurity		
Hazards of labour: anoxia and birth trauma	10	8
Congenital anomalies: bilateral atresia of meatus and Treacher Collins syndrome	10	8
Total	57	47
NEO-NATAL		
Kernicterus: Rhesus incompatibility	6	6

POST-NATAL		
Meningitis	5	4
Other acute infections	9	8
Toxic drugs: streptomycin ..	1	1
Total	15	13
UNKNOWN	40	34
GRAND TOTAL	118	100

Assessment

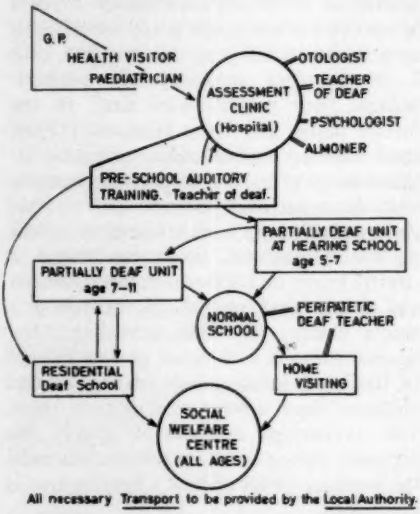
Having once found the children with a possible hearing loss the next problem is how to assess their hearing. Audiograms cannot be taken with any degree of accuracy until after the fourth year, but play techniques give a valuable indication of response to sound. It is extremely difficult to assess a child's hearing at one visit. The teachers of the deaf, who play a vital part in this assessment, often have to see the child on several occasions before giving a final verdict on his hearing or possible mental retardation. Many children whose deafness is due to maternal rubella have only a high-tone loss, and although they seem to babble and respond to sound intelligibly, their speech does not develop accurately or completely. The high-tone loss may not be recognised until an audiogram is done, and so children who develop speech late and who speak indistinctly should always have their hearing assessed. Unfortunately those with a high-tone loss do not get much help from an aid.

Assessment of hearing in very young children is a team problem, involving paediatrician, otologist, psychologist and teacher of the deaf. If there is any doubt about the child's hearing he should be fitted with an aid and given auditory training irrespective of the diagnosis. The youngest child so far fitted at Oxford is a baby of 4 months and many have been fitted as young as 9 months. Children very quickly learn to accommodate themselves to wearing a mould in their ear and often confirm that they are benefiting from their aid by demanding it. I would like to see two aids issued to more children so that they could make use of their residual hearing in both ears. After all, children with errors of refraction are given a pair of glasses, not monocles!

Training and Education

After assessment, the auditory training and future education of these children raise another set of problems, which vary in each locality. The decisions taken at this early age will vitally affect the child's whole future. Experimental work on this subject is being done in schools up and down the country and also abroad, particularly in Denmark.

It will give some idea of the factors involved if we consider the present handling of deaf children in Oxford (*see figure*).



Arrangements for deaf children at the Radcliffe Infirmary, Oxford.

An Assessment Clinic is held, usually once a week, in the Otological Department at the Radcliffe Infirmary, in a non-white-coated atmosphere. The Board of Governors employ two qualified teachers of the deaf attached to the hospital, and a third is employed by the Regional Hospital Board. One or both of the teachers attached to the hospital attend the weekly assessment clinic, together with the otologist, a psychologist and usually an almoner. The

registrar and housemen are also encouraged to attend, for it is of great value to the younger men to be taught something of the management of deaf children.

Children are referred to this clinic mainly by the paediatricians. Some come through the local general practitioners, some from the Ear, Nose and Throat Department, and some through the Medical Officers of Health. Lectures are given to health visitors on how to do elementary screening tests of the hearing of babies. Midwives also can attend the lectures.

Children attending the Assessment Clinic have already had a clinical examination and after assessment are handed over to the teacher of the deaf. She arranges to see them frequently, to assess their capabilities further and give their mothers help and advice. Parents can play a vital part in carrying on the continuity of auditory training. To make speech and sound more often available to the children, parents are advised to have their wireless and television sets at home looped for sound; this is a simple, inexpensive procedure and is of great help to children with loop aids. It might be of inestimable benefit if local authorities could arrange for each home with a severely handicapped child to be fitted with a loop amplifier.

The children are often admitted to the E.N.T. Department for assessment of residual hearing. The children's ward there is looped, and the Sister has four loop amplifying aids for the use of these children or any others whose hearing gives rise to suspicion while they are in hospital.

The follow-up and early auditory training of children is done either at the hospital or at the Social Centre for the Deaf. There is a classroom looped for sound in both places and the hospital teachers undertake the children's training. When the children are a little older they can attend a nursery school and continue to be taught by the same teacher of the deaf; at this school they will mingle with hearing and speaking children. The teachers also visit the homes of some children to give auditory training, especially out in the country or where domestic problems arise.

The local Education Authorities are most helpful and co-operative over this work. They arrange transport for the children and their mothers to attend the clinics, and also provide transport and lunch for children attending the nursery school.

From the ages of 5–7 years, when more formal education begins, children are admitted to a deaf unit attached to a normal hearing school in the town. There are two of these units and the local Education Authorities employ two teachers of the deaf. If, after passing through the first unit, a child is unable to take his place among the hearing children in the normal school, he will go on to the second unit (7–10 years of age). The children in these units have their lessons with the teachers of the deaf but mix for play, meals and certain lessons, such as handwork, with the hearing and speaking children. This integration with their contemporaries can be of very great value to handicapped children, but its success may depend on the way the headmaster or mistress explains the handicap of a hearing loss to the rest of the school. The work of these deaf units is still experimental. Partially deaf units are being tried in many parts of the country, often with great success.

When the children go on to the junior schools the senior teacher of the deaf visits them and continues to watch over their progress. This follow-up of the children is most important. So far there has been an unfailing supply of candidates seeking admission to the deaf units, and there has been a noticeable improvement in the confidence of the children who, while living at home, attend the units and mix with normal children.

The problems of the deaf and hard of hearing do not stop when school days are over, and their rehabilitation continues to be the responsibility of the medical profession, the teachers of the deaf, missioners and the general public. In Oxford we have

a Social Centre for people of all ages who have a hearing loss. The totally deaf, the partially deaf and the parents of deaf children all use the centre for rehabilitation, further education and social activities. Through it the parents and everyone in any way connected with a handicapped child can co-operate and continue their work for that child after he has left school and taken up employment. They can help him to adapt himself to his new environment and can watch and encourage him throughout his life.

Much work remains to be done to find the best methods of training and educating children with a hearing loss, and many people are willing to help. Lasting help for these severely handicapped children calls for continuous co-operation between all those concerned with their welfare.

SUMMARY

A child with normal hearing learns to imitate sounds and interpret them during the first 3 years of his life. Most so-called deaf babies have some residual hearing; if this can be trained during those early critical years the child will have his best chance of learning speech.

In order to find these handicapped babies one should know which must be considered 'at risk' or suspect, because they or their parents have been exposed to the known pre-natal, neo-natal or post-natal causes of deafness.

Since it is 14 times as common to find deafness among the 'at risk' children than among normal children, a register of these 'at risk' babies should be kept at all large hospitals. All 'at risk' babies should have their hearing tested frequently until the presence or absence of hearing loss is confirmed.

A hearing defect must also be suspected in backward children and those who develop speech late.

Children with bilateral congenital atresia of the meatus should be operated on between 2 and 3 years.

The problem of training and educating these children vary with each locality, but decisions taken before the age of 3 vitally affect the child's whole future.

RÉSUMÉ

Troubles de l'audition et de la parole durant l'enfance.

Un enfant dont l'audition est normale apprend à imiter les sons et à les interpréter au cours des trois premières années de sa vie. La plupart des bébés que l'on dit sourds gardent en fait quelque résidu d'audition; s'il est possible d'exercer ce résidu pendant ces premières années critiques, l'enfant tiendra sa plus belle chance pour apprendre à parler.

Afin de reconnaître ces jeunes enfants handicapés, il faut d'abord savoir lesquels d'entr'eux doivent être considérés comme étant 'en danger' ou suspects parce qu'eux mêmes ou leurs parents ont été exposés à des causes connues prénatales, néonatales ou postnatales de surdité.

Etant donné qu'il est 14 fois plus courant de trouver des enfants sourds parmi les enfants du groupe ci-dessus que parmi les enfants normaux, il faudrait faire procéder, dans tous les grands hôpitaux, au recensement de ces bébés 'en danger'.

Il faudrait que tous les bébés 'en danger' soient soumis à des tests fréquents et répétés jusqu'à ce que l'existence ou le défaut d'audition soient confirmés.

On doit également pressentir un vice de l'audition chez les enfants retardés et ceux qui se sont mis à parler tard.

Les enfants atteints d'atrésie congénitale bilatérale devraient être opérés entre 2 et 3 ans.

Former et instruire ces enfants sont des problèmes qui varient d'un lieu à l'autre, mais les décisions prisent avant l'âge de 3 ans pèsent de façon vitale sur l'avenir tout entier de l'enfant.

ZUSAMMENFASSUNG

Gehör- und Sprachdefekte des Kindesalters.

Ein Kind mit normalem Gehör lernt während der drei ersten Lebensjahre, die Laute nachahmen und deuten. Die meisten sogenannten tauben Babies haben noch einen Rest des Hörvermögens; wenn es möglich ist, diesen in den ersten kritischen Jahren auszuüben, wird das Kind seine beste Gelegenheit finden, sprechen zu lernen.

Da es 14 Mal häufiger ist, taube Kinder unter den Kindern der oben erwähnten Gruppe als unter normalen Kindern anzutreffen, müsste man in allen grossen Krankenhäusern diese Fälle von 'bedrohten' Babies registrieren.

Alle bedrohten Babies müssten häufig untersucht werden, bis man normales Hörvermögen oder Gehördefekt bewiesen hat.

Eine Störung des Hörvermögens muss man auch bei zurückgebliebenen Kindern oder bei solchen, die spät zu sprechen angefangen haben, verdächtigen.

Kinder mit doppelter angeborener Atresie müssten zwischen 2 und 3 Jahren operiert werden.

Das Problem der Bildung und des Unterrichts dieser Kinder wechselt von einem Ort zum andern aber die vor dem Alter von 3 Jahren genommenen Entscheidungen haben eine lebenswichtige Wirkung auf die ganze Zukunft des Kindes.

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Hearing and Speech Disorders

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THERE are close connections between hearing disorders and disorders of speech, and it is convenient to consider the two together. For, whether one works primarily in the field of deafness or in that of speech disorders as such, one will sooner or later meet a case involving a difficult differential diagnosis, where a knowledge of both fields is essential and there is a need for close liaison between workers in each. The problems of diagnosis and treatment are closely related in these cases, and, though treatment should always be preceded by diagnosis, this may present grave difficulties. It may be hard to make a diagnosis and just as difficult to arrange proper treatment. Often one may have to begin treatment before one can find out the child's true condition.

Most cases of hearing disorders are simple and straightforward, and in these there is little difficulty in diagnosing the deafness and assessing its degree. It is easy to assess a child's deafness accurately, provided he has a mental age of 3 years or more and is reasonably co-operative. The vast majority of children will co-operate if the adult demanding co-operation understands child management, particularly the management of children with communication difficulties. It is necessary to know something about hearing and deafness, something about the instruments to be used, and a great deal about children. Too often the first and second conditions are fulfilled but not the third. Too often unscientific information comes from scientific instruments. The person measuring a child's hearing must be skilled in using scientific instruments in the play way necessary with children, and particularly in instructing and gaining responses from the child who cannot communicate in the normal way.

Below a mental age of 3 years, one cannot expect the co-operation necessary for an accurate assessment of hearing loss, although one can make remarkably good estimates from the use of distracting sounds, from the child's history, and from the parents' reports.

My experience of speech disorders not due to deafness is limited, but the great majority of such cases are probably also straightforward. However, in many of them diagnosis is difficult because the child will not co-operate or is too immature, or because of an abnormal behaviour pattern. The investigation of these children requires knowledge of many different disciplines, a knowledge of both hearing and speech disorders, and the close co-operation of paediatricians, otologists, neurologists, psychiatrists, psychologists, teachers and parents.

The present demand for early awareness of abnormalities and the consequent demand for earlier and correct diagnosis carry disadvantages as well as advantages. It becomes increasingly difficult to make an accurate diagnosis, but time is on one's side for both diagnosis and treatment.

Clinical Responses and Possible Causes

Clinically, the problem may present itself as follows:

1. *Complete lack of response to sound*
   Possible condition:
   i. Severe deafness.
   ii. In a very young baby, moderate deafness.
   iii. Severe mental retardation or general immaturity.

iv. Psychological withdrawal.
v. An aphasic or dysphasic condition.
vi. A physical inability to make the normal responses.

2. *Lack of response to some sounds or a variable response*
Possible condition:
i. Partial deafness.
ii. Variable deafness.
iii. Mental retardation or immaturity.
iv. Psychological withdrawal (responses only when great interest is aroused).
v. Lack of attention.
vi. An aphasic or dysphasic condition.

3. *Normal response of awareness to all sounds but delayed or defective speech*
Possible condition:
i. Normal hearing.
ii. High-tone loss with normal or near normal low-tone hearing.
iii. Severe low-tone loss with normal or near normal high-tone hearing.
iv. An aphasic or dysphasic condition.
v. Developmental delay.

This third clinical picture is an important one, for the children in subgroups ii, iii and iv have often been declared mentally backward, and, since any of these disturbances may be accompanied by abnormal behaviour, it is all too easy to put these children into mental institutions. It is just as wrong to assess a child with a difficult behaviour pattern as deaf because he has not responded to sounds, and think that his abnormal behaviour pattern is due to deafness. The hyperactive child who rushes round the room is by no means an easy case to sort out but is nevertheless entitled to full investigation. The extremely withdrawn child, though not so wearing on the nerves, may present just as difficult a diagnostic problem. Both may require long-term investigation.

All children with speech defects and all children thought to be mentally retarded should be given a hearing test by people who understand this technique adequately. This last point cannot be stressed too much, for the more subtle forms of hearing impairment may be missed by people insufficiently trained for this work.

Many babies are now being seen in the first year of life, and most will respond in the expected way. Unfortunately, though a great deal is known about the way in which many babies react at various stages, too little is known about the deviations, and there seem to be many. It is not enough to see a baby and try to assess his hearing level; one must go on until one is sure of the results. In various parts of the country 'at risk' babies are now being sought out and examined. These studies are likely to throw up many babies with central nervous disorders which so complicate the expected reactions as to make assessment a long and difficult task. Such children may be unable to make the normal responses or may move about so continuously that it is difficult to decide what is and what is not a response to a stimulus. Many babies are so strongly visually attracted to people that they do not readily respond to distracting sounds. In many of these cases, testing in a light sleep gives readily identifiable responses to sound stimuli. Such responses have been obtained from many babies who gave no response at all while awake.

Assessment of Aphasic and Dysphasic Children

The aphasic child can rarely be assessed before the age at which a normal child should have developed a reasonable level of speech. If he is sufficiently co-operative and mature it is a relatively easy task to assess his hearing and mental levels. If these are within normal limits and the child's background is normal, and still he cannot speak, one must consider the possibility of an aphasic condition. Dysphasic conditions take much longer to show themselves and are much more difficult to establish, because it is only after a child has been in a learning situation for a considerable time that one can reasonably consider that delayed understanding or speech is due to his condition. By the time one begins to think of the possibility of an aphasic or dysphasic condition, the child has usually become so disturbed that few of these tests can be done readily. Only after prolonged observation at home or in a unit such as the one at Belmont Hospital can such a diagnosis be made.

The picture often seen, therefore, is that of a disturbed child. He may be hyperactive, extremely withdrawn, or tied to his mother, reasonably controlled and happy at home but not willing to do anything for a stranger.

In all cases one should try to make a play situation in which the child is likely to co-operate, and this situation must never depend on speech communication. If this kind of situation cannot be established, accurate diagnosis will be impossible. With such children, objective tests of hearing, which in theory should provide the answers, do not seem to work. Once the child begins to play, observations can more easily be made, and with more and more co-operation tests may be applied and conclusions drawn. Even if one finds a less than normal response in one area, one should be quite sure about other areas. A child could be partly deaf, somewhat dull and also dysphasic.

The combination of partial deafness and dysphasia was clearly demonstrated in the following case:

CASE-RECORD

Boy, born January 26, 1950.

History

Normal pregnancy. Normal birth, good colour. Operation for pyloric stenosis at 19 days. Whooping-cough at 3 yr. Chicken-pox at 4 yr. No family history of deafness. Sat up at 10 mos. Walked at 1 yr. 10 mos. No speech development. Deafness was suspected at 2½ yr.

At 3 yr.: In free field situation: responded to his name at 6 ft., moderate voice; responded to low tones (250 c/s, 500 c/s) at 12 ft.; responded to 1000 c/s tone close to his left ear. Recommended for nursery class for deaf children.

Report from school after 6 mos.: Did not mix with other children; would sometimes try to imitate them but did not play with them. Wandered idly staring at things, sometimes smiled to himself. Could not use simple gestures for communication. Would imitate voice but could not put speech sounds together. Would hit his head with his hand while walking about. Not certain that he has a hearing defect. The boy made several visits to the clinic, but showed very little co-operation. He would never leave his mother.

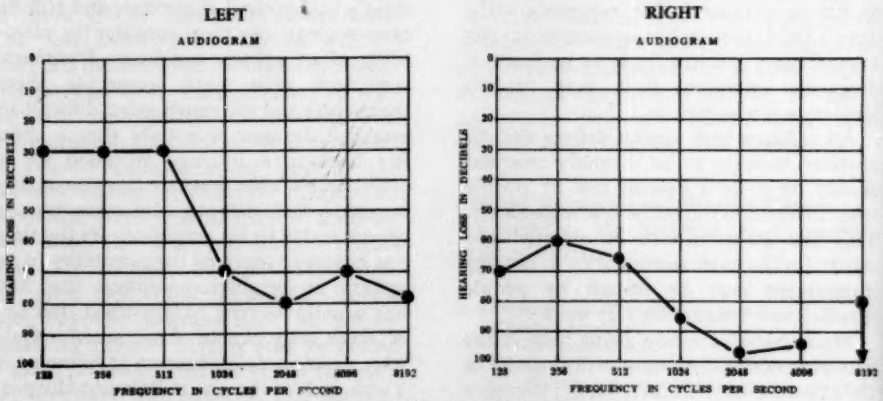
At 4 yr.: Aggressive and self-willed, but his general retardation seemed to be due to temperamental difficulties rather than general mental retardation.

At 4½ yr.: A pure-tone audiogram was obtained (see Figure). Not much to report until 8 yr. old. Not making expected progress in school.

Tests of Intellectual Ability

Wechsler Intelligence Scale for Children at 8 yr. 9 mos : I.Q. (non-verbal) 113. Sub-test scores: 12, 9, 18, 15, 7. His general behaviour in concrete, non-verbal situations agreed with the result of high-average general ability (non-verbal). He could lip-read only about 20/30 words and a few simple phrases. His ability to read and write were at an equally low level. This level of attainment in communication, in a school with generally high levels of attainment, was not commensurate with his innate ability or hearing level, and therefore a dysphasic condition must be suspected. A child with this level of hearing and ability should, by this age, have been speaking, hearing, reading and writing well, with perhaps a slight speech defect.

It was not until he had been in a learning situation for some time that one could diagnose a possible dysphasic condition. It is important to establish this possibility early so as not to expect too much of him and thereby add to his difficulties.



Pure-tone audiogram at 4½ years.

Placement and Prognosis

The correct placement of such a child is difficult, for there are few places available in this country for a condition that may not be so uncommon as is generally believed. The correct treatment is not obvious and the educational prognosis a bleak one.

Discussion

One must be very careful how one interprets mental-test scores from children with communication disorders. Some tests contain items which, though not verbal in themselves, nevertheless demand verbal experience in realising their significance. One must therefore consider on whom the norms of the items were established and whether the estimate of intellectual level is valid or not.

The child who is withdrawn and generally not quite with one is the most difficult of all to assess, and, so far as my experience is concerned, he also has the worst prognosis. One cannot obtain the necessary rapport to test for anything accurately, and one is never quite sure whether lack of communication is caused by a psychological withdrawal, or the withdrawal is a result of the inability to communicate. In such cases one often gets a low score on an intelligence test, but if the child has paid little attention to its environment this may mean very little.

All too frequently the diagnosis can only come out of the way in which one treats the child. Children with communication disorders not due to deafness have had the least attention from the educational system. There is undoubtedly a need for many more educational centres for children who are different, many of whom present such a disorder. Into such establishments, day or residential, these children should go to live a reasonable life where normal modes of communication may be at a discount and they learn to live with their handicap, while at the same time a diagnosis can be made in order to help them. They may never reach any useful level of ability to communicate and yet may learn, in a concrete situation, to carry out highly skilled procedures. Such children may have to mark time in an interesting and stimulating environment, until they are old enough to learn a skilled occupation which will enable them to live a useful life, complete except for normal communication.

These cases are not common but may be seen sporadically throughout the country. There is a need for centralisation of our knowledge of such cases, in order to work out the best kind of training for them in childhood and particularly for when they are about to be launched into adult life.

SUMMARY

Many children present a complex picture, of which the outstanding symptom is lack of response to sound or a varying response to sound. Many different conditions in children may give rise to this superficial picture of hearing defect, and all the possible causes have to be differentiated. This problem of differentiation is discussed in detail.

In some cases children may be thought to hear normally and yet have an impairment of hearing or of speech-perception. It is important to know of all the possible conditions that might be influencing the situation, so as to make an accurate diagnosis as early as possible.

RÉSUMÉ

Les troubles de l'audition et de la parole.

De mombreux enfants présentent un tableau complexe dont le symptôme principal est l'absence de réponse au son ou une réponse variable au son. De nombreuses maladies peuvent donner lieu, chez l'enfant, à ce tableau superficiel des troubles de l'audition et les causes possibles doivent être reconnues. Ce problème diagnostique est discuté en détail.

Dans certains cas, on considère que les enfants entendent normalement, ils peuvent cependant avoir une atteinte de l'audition ou de la perception de la parole. Il est important de connaître tous les états qui peuvent influencer cette situation afin de faire un diagnostic exact aussi précoce que possible.

ZUSAMMENFASSUNG

Gehör- und Sprachstörungen

Viele Kinder zeigen ein verwickeltes Bild, dessen Hauptsymptom Ausfall der Antwort auf den Klang oder eine veränderliche Antwort auf den Klang ist. Viele Krankheiten können bei Kindern dieses oberflächliche Bild der Gehörstörungen geben und es ist nötig, alle möglichen Ursachen zu erkennen. Dieses diagnostische Problem wird gründlich besprochen.

In gewissen Fällen nimmt man an, dass die Kinder normal hören. Sie können aber an einer Störung der Gehörfähigkeit oder der Wahrnehmung der Sprache leiden. Es ist nötig, alle Bedingungen, die einen Einfluss auf diesen Zustand ausüben können, zu kennen, um eine genaue Diagnose so rasch wie möglich zu stellen.

A Description and Classification of Common Speech Disorders Associated With Cerebral Palsy

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MANY children with cerebral palsy have defective speech. In different series different percentages of patients with cerebral palsy who have speech defects are reported, depending on the criteria of diagnosis used, the selection of patients and the care with which they have been examined. Between 30 and 70 per cent of children with cerebral palsy were considered to have significant speech disorders in the four series reported by Dunsdon (1952), Ingram (1955), Skatvedt (1958) and Illingworth (1958).

In this paper we present an account of the commoner disorders of speech encountered among children with cerebral palsy seen in the Speech Clinic of the Royal Hospital for Sick Children, Edinburgh, and in the in-patient and out-patient work of the Scottish Council for the Care of Spastics in Edinburgh.

We have analysed the results of our examinations of 258 children in order to get some idea of the frequency with which different types of speech disorder occur in the common forms of cerebral palsy. The numbers of children with the less common types of cerebral palsy (bilateral hemiplegia, ataxic diplegia and ataxia) were not considered sufficient to merit a statistical presentation of their findings. Since the children we studied were highly preselected, our findings must not be taken as representative of those in the general population of children with cerebral palsy.

Terminology and Classification of Cerebral Palsy

Since there is much confusion because of the varying usage of descriptive terms and the employment of different classifications of cerebral palsy and speech defects, some account must be given of those we use.

The terminology and classification used are those suggested by Balf and Ingram (1955). The term 'cerebral palsy' is used to describe impairment of motor function of one or more limbs due to paresis, involuntary movement or inco-ordination which is attributable to nonprogressive brain disease present at the time of birth or shortly after.

Children are classified according to their neurological diagnosis and are further categorised according to the nature, severity and extent of their motor handicaps (Table I).

Further classification takes account of the extent and severity of the impairment of function in the limbs. The extent of the significant loss of limb function, involving one limb only, both lower limbs, three limbs or four limbs may be indicated most easily by using the terms monoplegic, paraplegic, triplegic or tetraplegic. Some clinicians have been worried lest these terms should be misunderstood as comprising medical diagnoses, but their use in a purely descriptive sense seems logical.

TABLE I—CLASSIFICATION OF CASES OF CEREBRAL PALSY IN CHILDHOOD
(Balf and Ingram 1955)

Neurological Diagnosis	*Distribution*	*Severity*
Hemiplegia	Right Left	Mild Moderately severe Severe
Bilateral hemiplegia		Mild Moderately severe Severe
Diplegia:		
Hypotonic	Paraplegic	Mild
Dystonic	Triplegic	Moderately severe
Rigid or spastic	Tetraplegic	Severe
Ataxic diplegia:		
Hypotonic	Paraplegic	Mild
Spastic	Triplegic	Moderately severe
With contracture	Tetraplegic	Severe
Ataxia:		
Cerebellar	Mainly unilateral	Mild
Vestibular	Bilateral	Moderately severe Severe
Dyskinesia:		
Dystonic	Monoplegic	Mild
Choreoid	Hemiplegic	Moderately severe
Athetoid	Triplegic	Severe
Tension	Tetraplegic	
Tremor		
Other types		

The severity of the cerebral palsy is assessed differently according to the type from which the individual child suffers. If he has hemiplegia, for example, the disability of the upper limb is the criterion of assessment, since this is usually the most severely affected part; while in diplegia the degree of disability in the lower limbs is the major consideration when severity is being estimated.

Further classification by aetiology is sometimes useful when speech defects are considered. Acquired right hemiplegia in a child of 3 years or more is much more likely to be associated with specific disturbances of language, for example, than is a congenital right hemiplegia. A child suffering from post-kernicteric dyskinesia is much more likely to be deaf than one whose involuntary movements are the result of perinatal hypoxia.

Hemiplegia is a unilateral paresis of voluntary movement which is usually more severe in the upper limb than the lower and is usually accompanied by spasticity and retardation of growth in the affected parts. Contracture, involuntary movements of athetoid type and vasomotor changes are usually seen also, and sensory impairment is common.

Bilateral or double hemiplegia is a paresis of all four limbs in which the upper limbs are more affected than the lower. With the paresis there is usually associated a spastic increase of tone in all limbs and some paresis of the bulbar musculature. The children are usually helpless and the majority are mentally defective and epileptic. Associated developmental malformations are common.

The term '*diplegia*' describes a more or less symmetrical paresis, presumably of cerebral origin, affecting the lower limbs more severely than the upper, and dating from birth or shortly afterwards. There may be generalised hypotonia, but it is more usual to find rigidity or spasticity alone or in combination. Mental defect, epilepsy and strabismus are commonly present, but bulbar paresis is rarely severe.

In *ataxic diplegia* there is paresis, more severe in the lower limbs than the upper, and ataxia of cerebellar type. There may be hypotonia in the affected limbs, but more often there is spasticity. Most of the children are mentally retarded and many are epileptic. A positive family history of ataxia or spastic paresis is often obtained.

Ataxic children show inco-ordination of movement and impaired balance as their presenting clinical features. In some children the unsteadiness is more to one side than the other. There is usually associated hypotonia in the affected limbs, though the tendon jerks may not be significantly depressed.

The term *dyskinesia* is used to describe children in whom involuntary movements rather than actual paresis are the major cause of physical handicap. The movements may be choreoid, athetoid, tremulous or dystonic (in the sense in which this term was used by Jakob 1925 and Herz 1944). In addition to actual involuntary movement, sudden involuntary increases in muscular tonus, known as 'tension', are often found in dystonic children.

Children in '*Other Categories*' represent those with mixed clinical pictures, most commonly combinations of hemiplegia, diplegia or ataxic diplegia with dyskinesia, and also those children who are still too young for an accurate diagnosis to be possible.

TABLE II—CLASSIFICATION OF THE COMMON SPEECH DISORDERS IN CHILDHOOD
(Ingram 1959a)

1. Disorders of voicing (Dysphonia).

(*a*) With demonstrable disease of the larynx.
(*b*) Without demonstrable disease of the larynx.

2. Disorders of rhythm (Dysrhythmia).

(*a*) Clutter.
(*b*) Stammer or hesitation.

3. Disorders of articulation with demonstrable dysfunction of articulatory apparatus (Dysarthria).

(*a*) Due to neurological abnormalities.
Cerebral palsy.
Suprabulbar palsy.
Lower motor neurone lesions.

(*b*) Due to local abnormalities.

Jaws and Teeth ..	Hypomandibulosis.
	Other malocclusion.
Tongue	Tie.
	Tongue thrust.
Lips	Cleft lip (only).
	Other.
Palate	Cleft (with or without cleft lip).
	Other.
Pharynx	Large pharynx (palatal disproportion).
	Acquired disease.
Mixed.	

4. Disorders of articulation without demonstrable dysfunction of articulatory apparatus (Secondary Speech Disorders).

(*a*) Secondary to hearing defect.
(*b*) Secondary to mental retardation.
(*c*) Secondary to psychogenic disorders.
(*d*) Secondary to dysphasia due to brain damage.

5. The Developmental Speech Disorder Syndrome (Specific Developmental Speech Disorders).

(*a*) Involving language development and articulation.
(*b*) Involving articulation only.

6. Mixed Cases.

7. Unclassified and Other.

Terminology and Classification of Speech Disorders

We distinguish speech disorders according to the function of speech which is most severely disturbed. Firstly it must be decided if a child suffers from a defect of voicing, rhythm or articulation (Table II; Ingram 1959).

Further classification of speech disorders is by associated clinical findings. Thus abnormalities of articulation are classified according to whether or not they are the result of demonstrable abnormalities of the structure or function of the articulatory organs. When abnormalities can be demonstrated which explain the articulatory defects, speech is called '*dysarthric*'. When the articulatory organs are normal and abnormalities of articulation are associated with mental retardation, severe psychiatric disorders, hearing impairment or dysphasia, they are considered to be '*Secondary*' (Ingram 1959).

The severity of speech disorders is very difficult to gauge accurately, especially in the conditions of the outpatient clinic. We have adopted as our criteria of severity the intelligibility of speech to relatives and other people the child meets. Children with normal speech are assumed (somewhat rashly) to be intelligible to most people most of the time. Those with mild speech defects are consistently intelligible to adults who know them well, but not to all strangers. Children with moderately severe speech defects are consistently intelligible to siblings and parents but not to people who have no daily contact with them, while those with severe speech disorders are not intelligible even to these. When we have been unable to form any definite impression of the severity of the child's speech disorder by questioning parents and relatives we have relied on our own estimates.

Some explanation is required for the omission of a category of speech disorder in which the major disturbance is in the ability to use language. We believe that the most important manifestation of language difficulty before the age of 3 years is retarded speech development, not a clinical syndrome of difficulty in using words ('dysphasia') as in the adult. 'Congenital dysphasia' is often a useful theoretical concept. Whether it can be shown to exist in actual clinical practice is highly debatable. Whereas the inclusion of a category of congenital dysphasia is objectionable on theoretical grounds, a category of acquired aphasia, which would allow for the placing of older children who develop language disorders after a period of normal speech development, is not (Guttmann 1942). The number of such children is relatively small, however, and in the present simplified classification no allowance is made for them.

Dysphonia. Disorders of voicing account for no more than 4 per cent of children in most large series of children with speech defects. The only disorder of the voice which occurs at all commonly in the ordinary speech clinic is 'dysphonia', or chronic loss of voice. Various degrees of this may be recognised, but most of the children sound as if they were 'stage whispering'. The loss of voice is often assumed to be functional on very inadequate evidence. Dysphonia must not be confused with inspiratory speech, found frequently in children with dyskinesia.

Dysrhythmia. Speech dysrhythmia, or the involuntary interference with the normal rhythm of speech, is one of the commonest disorders of speech in childhood. A high proportion of otherwise normal children between the ages of 3 and 4 years hesitate or stammer for a period of 3 to 12 months. Such a transient disturbance of speech has been called 'clutter', by speech therapists.

Hesitation or stammer may be found also in children of school age or older, without detectable abnormalities of the nervous system. Many of them are ambidextrous, or show little lateralisation of handedness, visual-fieldedness or footedness. A high proportion have a family history of hesitation, stammer or other speech defect (Morley 1957).

The proportion of children who stammer or hesitate because of organic disease of

the nervous system is small, but it includes a relatively high proportion of the children whose speech disorder is due to cerebral palsy. Speech dysrhythmia is common in children suffering from dyskinesia, but it also occurs, less commonly, in those with hemiplegia, ataxic diplegia and ataxia.

Dysarthria is defective speech which is attributable to abnormalities in the form or function of the organs of articulation. Hare-lip and cleft-palate are typical structural abnormalities. Neurological disorders may cause paresis, inco-ordination or involuntary movement of the articulatory organs.

The abnormalities of function observed on careful examination of the mouth, pharynx and respiratory movements must be carefully correlated with the defects of articulation which occur. These are usually consistent and rather characteristic. For example, a paretic palate will fail to close off the nasopharynx during speech and there will be hypernasality, with abnormalities of all consonants except 'm', 'n', and 'η'. Paresis with spasticity of the lips may result in defective articulation of 'p', 'b', 'm', 'w', 'v', 'f', 'u', and 'o'. The analysis of the speech defects when there are severe involuntary movements of the articulatory organs is more difficult.

There is a tendency to regard all children with cerebral palsy who have defective speech as suffering from 'Dysarthria'. Yet simple retardation of speech development is a commoner cause of unintelligible speech. It is often forgotten that the definitive diagnosis of dysarthria depends on a demonstration of dysfunction of the articulatory organs sufficient to account for the observed defects of articulation. Dysarthria is much commoner in bilateral cerebral palsy than in hemiplegia.

Secondary speech disorders are those in which abnormalities of articulation are attributable to physical or psychological disorders not affecting the articulatory apparatus specifically. The majority of these cases show simple retardation of speech development. Severe deafness, mental retardation and major psychiatric disorders affecting speech are usually manifest in this way. Speech sounds are acquired late. In addition, acquisition of words and syntactical structure may be slow, and comprehension of speech may also be late in development. In not too severe high-tone deafness there is usually some general retardation of speech development, but selective impairment of the ability to acquire high frequency speech sounds is often obvious; 'f' and 's', for example, are often absent. Secondary speech disorders are commonly found in children with cerebral palsy. The most frequently observed abnormality is simple retardation of speech development, usually associated with mental retardation or defective hearing.

Specific Developmental Speech Disorders ('Developmental dysphasia' and related disorders). These comprise disorders of articulation which are not attributable to abnormalities of the articulatory apparatus or to other manifest disease. Children suffering from these conditions show unexpectedly slow development of intelligible speech, though in other respects their development is usually normal and their health is good (Ingram 1959). Specific developmental speech disorders are the commonest cause of abnormal speech in the general child population and so a proportion of children suffering from cerebral palsy are likely to be affected. Because an identical clinical picture may result from brain injury, certain diagnosis is extremely difficult and often impossible.

Mixed Cases are those in which more than one cause for defective speech is present—for example, children with cleft palate and deafness, which often results from the otitis media to which they are liable. A very high proportion of children with cerebral palsy and defective speech fall into this category. Children with choreoathetosis may show a mixture of dysarthria, dysrrhythmia and secondary speech disorder due in part to deafness and in part to mental retardation. The analysis and classification of the various causes of

defective speech is especially valuable in such complex cases. It becomes possible to assess the contribution that each abnormality is making to the total clinical picture of the speech defect. This in turn puts therapy on a more rational basis than is often the case.

The category of '*Unclassified and Other*' is used for children with defective speech in whom full diagnostic assessment has not been possible. It is useful, for example, for children whose co-operation in the testing of hearing is limited and in whom no definite assessment of hearing or mental ability can be made.

The Speech Defects found in Cerebral Palsy

The fact that different disorders of speech are so commonly associated makes their diagnosis in individual children with cerebral palsy difficult. The fact that language development is much retarded in a mentally defective child with cerebral palsy may be masked by his being severely dysarthric. An isolated defect of speech may often be due to a number of different causes. For example, inability to pronounce 's', 'th', 'l', or 'r' in a severely affected diplegic child may be due to simple retardation of speech development associated with mental retardation, or be attributable to dysarthria. Even a detailed examination may fail to elucidate the cause of all the speech defects encountered in children suffering from the various types of cerebral palsy. These will be considered in turn.

Speech Defects in Children with Hemiplegia

Normal speech is more commonly found in children with hemiplegia than in those with any other type of cerebral palsy. In a third of a series of hemiplegic children, mostly of school age, no defect of speech was found (Tables III and IV). In about 25 per cent the speech defect was considered to be mild and in 42 per cent it was moderate or severe. No significant difference was found in the proportion of right and left hemiplegic children with

TABLE III—TYPE OF SPEECH DEFECT IN 69 HEMIPLEGIC CHILDREN (46 WITH CONGENITAL AND 23 WITH ACQUIRED HEMIPLEGIA) BY INTELLIGENCE

I.Q.	*No speech disorder*	*Retardation only*	*Dysarthria only*	*Dysphasia only*	*Dysrhythmia*	*No speech*	*Not exclusive categories*				*Total*
							Retardation + other defects	*Dysarthria + other defects*	*Dysphasia + other defects*	*Dysrhythmia + other defects*	
90+ ..	12	2	0	2	1	0	3	2	0	2	21
70–90 ..	8	5	0	0	0	0	3	3	0	0	16
50–70 ..	3	11	0	0	1	0	5	4	6	6	23
—50 ..	0	8	0	0	0	1	0	0	0	0	9
Total ..	23	26	0	2	2	1	11	9	6	8	69
Approx. %	34	38	0	3	3	1	16	13	9	12	100

TABLE IV—SEVERITY OF SPEECH DEFECTS BY SIDE AND AETIOLOGY OF HEMIPLEGIA IN 69 PATIENTS

Aetiology	*Right Side*			*Left Side*			*Grand Total*
	Congenital	*Acquired*	*Total*	*Congenital*	*Acquired*	*Total*	
No speech defect ..	10	4	14	5	4	9	23
Mild defect	6	2	8	7	2	9	17
Moderate defect ..	6	5	11	3	1	4	15
Severe defect or no speech	4	3	7	5	2	7	14
Total	26	14	40	20	9	29	69

speech defects (Table IV). There was no statistically significant difference in the proportions of children with congenital and acquired hemiplegia who had speech defects. The proportion of children with speech defects was higher in those with acquired right hemiplegia than in those with acquired left hemiplegia or with right or left congenital hemiplegia. This is in accordance with the findings of Crothers and Paine (1959).

In hemiplegic children, the commonest disorder of speech was simple retardation of speech development. The acquisition of speech sounds, words and phrases was slowed. Retarded maturation was the only abnormality of speech in 18 of 46 children suffering from congenital hemiplegia and in 8 of 23 cases with acquired hemiplegia. The retardation was manifest in the slow acquisition of words, as well as speech sounds in the majority of cases. Retardation of speech development was associated with mental impairment in all but 2 of the congenital cases and in all of those with acquired hemiplegia (Table IV).

Dysarthria as an isolated defect of speech is uncommon in hemiplegics. Much more often it is associated with retardation of speech development or with dysrhythmia. In the present series, 6 of the 46 children with congenital and 3 of the 23 with acquired hemiplegia showed abnormalities of movement of the articulatory apparatus causing faulty articulation. In none was this dysarthria the only abnormality of speech. Dysarthria was no more frequent among children with acquired hemiplegia than among those with congenital hemiplegia, which was an unexpected finding.

Dysarthria slows the rate of utterance and causes incorrect articulation of many consonants. The consonants affected varied with the nature, distribution and severity of the involvement of the articulatory organs. Plosives, labiodentals and interdentals were most often abnormal. Hypernasality due to defective palatal movement is not common and occurred in only 3 of the 69 hemiplegic patients.

Dysrhythmia was present in 7 of the 46 children with congenital hemiplegia and in 3 of the 23 with acquired hemiplegia—approximately 14 per cent of all the hemiplegic children. It occurred as a single abnormality in 2 patients and in association with retardation of speech development, with dysarthria or with dysphasia, in the other 8. It was usually manifest as a combination of arrest or hesitation in the flow of speech with stammer. It is important to distinguish the temporary arrests of speech, caused by disrhythmic hesitation, from those caused by dysphasic difficulties.

True acquired dysphasia, manifest as an inability to use previously acquired words correctly, in order, at will, was present in 8 patients, 3 with congenital and 5 with acquired hemiplegia. In 2 patients it was the only abnormality of speech (Table IV). In 2 of the congenital cases, dysphasia was first observed after severe epileptic attacks. One of the dysphasic patients had an acquired hemiplegia on the left side and a family history of sinistrality.

Bilateral Hemiplegia. In bilateral hemiplegia, dysarthric speech defects are almost inevitable, for the bulbar muculature is involved. However, since most of the patients are severely mentally defective, the associated retardation of speech development usually conceals the extent of the paresis of the articulatory organs.

Most children have more or less severe feeding difficulties in infancy, which may involve both sucking and swallowing or swallowing only. There is usually an associated drooling, commonly exercerbated when teething begins. Many children regurgitate frequently an may inhale foodstuffs.

Evident compression of speech is usually very slow to appear if it comes at all. It may be impossible to determine whether an individual child's inattention to speech is the result of an intellectual handicap, conductive or central deafness, or inability to show signs of even limited comprehension. Speech development is usually rudimentary at best, and no more than a

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TABLE V—TYPES OF SPEECH DISORDER IN 122 DIPLEGIC PATIENTS BY EXTENT OF LIMB INVOLVEMENT

Extent of limb involvement	*No abnormality of speech*	*No speech*	*Dysarthria only*	*Retardation only*	*Dysrhythmia only*	*Dysphasia only*	*Not exclusive categories*				*Total*
							Dysarthria + other speech defects	*Retardation + other speech defects*	*Dysrhythmia + other speech defects*	*Dysphasia + other speech defects*	
Paraplegia	22	1	6	6	1	1	13	13	2	1	49
Triplegia	2	0	3	1	0	0	3	3	0	0	9
Tetraplegia	6	21	16	8	1	0	14	13	3	1	64
Total	30	22	25	15	2	1	30	29	5	2	122
Approx. %	24	18	20	12	2	1	24	23	4	2	100

few single words and rudimentary phrases are acquired. Articulation is grossly defective. There is usually marked nasal escape, and in many cases only the early acquired consonant sounds, such as *m*, *n*, and *g*, are distinguishable.

Speech Defects in Diplegia

A relatively high proportion of diplegic children have defective speech. It is commoner in those with the more extensive limb involvement than in those who are less severely paralysed.

Thus about half the paraplegics in the present series had apparently normal speech, but only a tenth of those with tetraplegia. In diplegia, as in hemiplegia, the commonest cause of abnormal speech is retardation of speech development. This was present as the only abnormality in 12 per cent of the diplegic children and in association with other causes of speech disorder in 23 per cent (Table V).

In addition to these cases, 22 children (18 per cent), most of them severely mentally retarded, had no more than one or two words of speech; 21 of them were tetraplegic. Retardation of speech development associated with gross mental defect appears to be the major cause of the failure to develop speech in these children but a high proportion had had feeding difficulties in infancy, with persistent drooling of saliva. In many of them paresis of the articulatory organs may have been a contributory cause.

In general, the children with the most severe and extensive paresis of the limbs showed the most defective intelligence and the greatest retardation of speech development. Hearing impairment was known to be a contributory cause of retardation of speech development in only 3 of the 122 diplegic children.

Dysarthria was commoner among diplegic than among hemiplegic children. It occurred more often in tetraplegic than in triplegic or paraplegic children. It was invariably accompanied by impairment of voluntary movements of the lips, tongue or palate, and in most of the severely affected cases all three organs were involved. A high proportion of children with severe dysarthria or no speech

TABLE VI—FREQUENCY OF DYSARTHRIA IN 49 DIPLEGIC PATIENTS WITH A HISTORY OF FEEDING DIFFICULTIES OR PERSISTENT DROOLING OF SALIVA IN INFANCY

	No. with feeding difficulties and/or drooling of saliva	*No. with no speech*	*No. with dysarthric speech*	*No. with speech not showing dysarthria*
Paraplegia	14	0	10	4
Triplegia	3	0	3	0
Tetraplegia	32	15	16	1
Total	49	15	29	5
Approx. %	100	30	60	10

development had a history of feeding difficulty in infancy or drooling (Table VI). Many of these had a positive sucking reflex.

Children of low intelligence seemed more likely to be dysarthric than those of fairly normal intelligence—perhaps because the more intelligent children are better able to compensate for their dysarthric difficulties.

The form of the dysarthric speech disturbance in diplegic children is characteristic. There is slowing of utterance and intonation, and stress patterns are unchanging and monotonous. There is often slight or moderate nasal escape because of palatal paresis, with resultant hypernasality and wastage of breath. Speech sounds are laboured; vowels are much better pronounced than consonants. The consonants which are defective vary with the distribution and severity of the neurological involvement of the articulatory apparatus.

Dysarthria occurred as the only abnormality of speech in about 20 per cent of the diplegic patients, and in association with other speech disorders in a further 24 per cent (approximately). It was most commonly associated with retardation of speech development.

It will be seen from Table V that relatively few children suffered only from significant dysrhythmia, dysphasia or dysphonia.

Speech Defects in Children with Ataxic Diplegia and Ataxia

Most children with ataxia and ataxic diplegia have defective speech. As with the other forms of cerebral palsy, the commonest disorder is a simple retardation of development. This may occur alone, but is commoner in association with dysarthria and other abnormalities, including dysrhythmia of a characteristic type.

The retardation of speech development is usually proportional to the degree of mental impairment, but it tends to be rather greater in ataxic children than in those with spastic paresis only. Like diplegic children, those with ataxic diplegia tend to show rather greater retardation of speech development if they are tetraplegic than if they are triplegic or paraplegic.

Dysarthric children often show a very striking inco-ordination between the movements of the lips, tongue and palate. This can easily be demonstrated by asking a child alternatively to whistle and to open his mouth and protrude his tongue. Children with ataxic diplegia show similar inco-ordination of voluntary lip, tongue and palatal movements, but in addition many of those with tetraplegic involvement show paresis of these organs, like diplegic children.

The slowness and ataxia of movements of the articulatory organs in ataxic disorders causes abnormalities of articulation which may be very similar to those resulting from the slowness and weakness of their movement in children with extensive diplegia. In ataxic disorders the abnormalities of pronunciation tend to be less consistent, however, and it is noticeable that sound sequences requiring major adjustments of lip and tongue position are particularly badly spoken. It is not uncommon to observe excessive nasal escape while an ataxic child talks, even though the movements of his palate appear to be full in range on examination. Probably the movement aimed at closing off the nasapharynx during speech takes place too slowly to be completed.

Dysrhythmia and associated abnormalities of intonation and stress, which are very difficult to analyse, occur in a high proportion of ataxic children. The dysrhythmia consists of an irregular division of phrases into segments of different lengths. At the beginning of the phrase there is a tendency for speech to accelerate, and then at about its middle to decelerate. With the acceleration the pitch rises and with the deceleration it falls. The resultant 'scanning' effect is characteristic of the speech of ataxic children.

Speech Defects in Children with Dyskinesia

Only a small proportion of dyskinetic children speak normally; the majority have

TABLE VII—TYPE AND SEVERITY OF SPEECH DEFECTS IN 67 DYSKINETIC CHILDREN BY AETIOLOGY AND INTELLIGENCE

I.Q.	Total	Impaired hearing	Retarded only	Dysarthria only	Dysrhythmia	Not exclusive categories			No speech	Severity		
						Retarded+	Dysarthria+	Dysrhythmia+		Mild	Mod.	Severe
Rhesus Incompatibility												
90–110 ..	5	4	0	1	0	2	4	4	0	1	3	1
70–90 ..	4	3	0	0	2	1	2	2	0	2	2	0
50–70 ..	12	8	2	0	0	6	2	2	4	0	2	6
−50	2	2	0	0	0	0	4	0	2	0	0	0
Total	23	17	2	1	2	9	12	8	6	3	7	7
Approx. % ..	100	72	8	4	8	39	52	35	26	14	30	30
Birth Injury												
90–110 ..	16	0	0	4	1	2	11	10	0	8	6	2
70–90 ..	11	1	0	2	1	1	7	7	1	3	4	3
50–70 ..	15	3	1	0	0	8	9	8	4	0	3	8
−50	2	0	0	1	0	1	1	1	0	1	0	1
Total	44	4	1	7	2	12	28	26	5	12	13	14
Approx. % ..	100	9	2	16	5	27	63	59	12	27	29	31
All patients ..	67	21	3	8	4	21	40	34	11	15	20	21
Approx. % ..	100	31	4	12	6	31	59	51	16	22	30	31

complex speech defects due to a number of different causes in each case. The 67 children with dyskinesia studied here all had defective speech. About 78 per cent had speech defects or a virtual absence of speech due to multiple causes. Only in about 22 per cent were the disorders thought to be entirely the result of dysarthria, retardation of speech development or dysrhythmia.

There is a smaller proportion of mentally retarded children among those with dyskinesia than among those with other types of predominantly bilateral cerebral palsy. The prevalence of retarded speech development from this cause is therefore lower among the dyskinetic children, but retardation of speech due to impairment of hearing is much more frequent in them than in other categories. Impairment of hearing and mental retardation are found more often among post-kernicteric patients with dyskinesia than among those whose involuntary movements are apparently the result of 'birth injury'. It is hardly surprising, therefore, to find that retarded speech development as the only abnormality of speech or in association with other speech defects was commoner in the children who had suffered from rhesus incompatibility (Table VII).

Dysarthria occurred as the only abnormality of speech in 8 patients. It was combined with dysrhythmia in 18, with dysrhythmia and retardation of speech development in 14 and with retardation of speech only in 6. Thus about 70 per cent of the 67 children showed dysarthria. The abnormalities of voluntary movements of the lips, tongue and palate observed in these dyskinetic children varied widely. In some, in the middle of a voluntary act, there would be an apparent arrest of the movement of tongue and palate, which appeared to be equivalent to the intermittent state of tension which occurred frequently in the limb musculature. Practically all the patients showed involuntary movements of the face, tongue, palate and often other parts of the body whenever the child attempted to move the articulatory organs to command. Thus a request to severely affected children to 'smile' would often result in intense bilateral facial grimacing, protrusion of the tongue and swallowing. The reproducibility of the involuntary movements which occurred varied widely from patient to patient, however, and from time to time in the same patient. The speech sounds which were defective varied similarly in dysarthric patients with dyskinesia. *Th*, *r*, *l*, *sh* and *p* were particularly liable to be wrongly articulated, however, especially when they occurred in combination or in close proximity to other consonants. Speech

tended to be slow and extremely laboured in children in whom this effect of dysarthria could be analysed.

In only 6 per cent of the dyskinetic patients studied did dysrhythmia appear to be the only abnormality of speech. It was associated with dysarthria in 27 per cent and with dysarthria and retardation of speech development in 21 per cent. Thus irregularities of rhythm were present in more than half the children studied (Table VII). The irregularities were almost invariably due to the irregular involuntary action of the respiratory muscles, which did not move synergistically with those concerned in articulation. In consequence, speech might be suddenly arrested, or a sudden unexpected involuntary inspiration would cause inspiratory speech. Occasionally, however, transient arrest of speech appeared to be due to obstruction of the air stream at a higher level, either in the larynx or the glottis. Irregularities of rhythm of speech of any severity are inevitably associated with a lack of normal intonation and of normal stress patterns. Together with dysarthria these disturbances often result in speech being almost entirely incomprehensible, even in the most intelligent child with good use of language.

Dysphonia is found occasionally in dyskinetic patients. Physical abnormalities of the vocal cords are not always obvious, but it seems likely that at times during speech the cords may be involuntarily fully abducted and this may cause loss of voice. This might explain the intermittency of the dysphonia in many of the dyskinetic patients who show it. In the present series of 67 dyskinetic patients, dysphonia was observed in 5.

The frequency with which various types of speech disorders are combined in individual patients with dyskinesia further increases the difficulties of their diagnosis and treatment. In the series studied, three-quarters of the dyskinetic patients had virtually no speech, or had complex speech defects comprising combinations of dysarthria, dysrhythmia and retarded speech development.

Discussion

Before there can be logical, usefully directed speech therapy there must be accurate diagnosis of speech disorders. This requires careful history taking and detailed examination of the patient, particularly his articulatory organs, his hearing and his nervous system. Abnormalities of the movements of his articulatory organs must be correlated with his articulatory defects, and the abnormalities of his speech must be considered in the light of the results of other medical and psychological examinations. If the results of examination are to be interpreted readily it is necessary to have an acceptable terminology and classification of speech disorders, comprehending anatomical, neurological and psychological aspects as well as those of speech. Yet in many clinics dealing with children with cerebral palsy the diagnosis as well as the treatment of speech disorders is not jointly discussed and planned but is deputed to speech therapists working alone. Often there is neither consistent use of terms nor an acceptable classification of the speech defects met and treated.

Here we have tried to apply the methods of study and classification of speech disorders used in the Royal Hospital for Sick Children, Edinburgh, to patients with cerebral palsy. As was expected, in a high proportion of cases the speech defects fell into more than one category of the classification. Nevertheless, we believe that, because it indicates the nature of the speech disturbances present, the classification is useful to the therapist. For example, a patient may be described as suffering from 'moderately severe dyskinesia with retarded speech development due to deafness, mental retardation, dysarthria and speech dysrhythmia'. Such a description may be cumbersome but does give more information to the therapist than one like 'severely defective speech', or 'poorly developed speech', to quote too commonly used descriptions.

The establishing of an acceptable classification of speech disorders is important from another point of view. At

present, many different forms of speech therapy are given almost indiscriminately to a large number of patients with different disorders of speech. Yet few clinicians caring for children with cerebral palsy appear to be at all impressed by the results of speech therapy. Until some acceptable method of classifying speech disorders in some detail is evolved, it is not going to be possible to make any controlled study of the effects of therapy.

No more is claimed for the present classification than that it enables most forms of defective speech in children with cerebral palsy to be described. We hope that its presentation may stimulate the production of improved classifications which should be more comprehensive and less cumbersome.

Acknowledgements: We are grateful to the Scottish Council for the Care of Spastics, who encouraged and supported this research project, and particularly to Mr. G. A. Pollock and Dr. J. A. L. Naughton for allowing us free access to their detailed clinical reports. We also wish to thank Prof. R. W. B. Ellis for his stimulation and advice.

SUMMARY

The speech defects of children with cerebral palsy are often neglected by doctors when attempting to make comprehensive assessments of their handicaps. Speech therapists are then left with the double burden of diagnosis and treatment.

Classifications of cerebral palsy and of speech defects in childhood which have been found useful in Edinburgh clinics are described. Using them, an attempt is made to define and give accounts of the common disorders of speech encountered in children suffering from the various types of cerebral palsy. The speech defects of 258 patients referred to the Edinburgh Clinic of the Scottish Council for the Care of Spastics are described.

In hemiplegic patients, retardation of speech development secondary to mental backwardness is a much more frequent cause of defective speech than is dysarthria, dysphasia or dysrhythmia. Children suffering from cerebral palsy which affects the limbs bilaterally are often more dysarthric than the hemiplegics, but retardation of speech development secondary to mental retardation or impaired hearing is still more important. A high proportion of patients with bilateral cerebral palsy have speech defects due to a number of contributory causes. It is important to recognise this fact when attempting to diagnose and treat abnormalities of speech in cases of cerebral palsy.

RÉSUMÉ

Description et classification de troubles habituels de la parole associés à l'infirmité motrice cérébrale.

Les troubles de la parole chez les enfants souffrant d'infirmité motrice cérébrale sont souvent négligés par les médecins lorsqu'ils essayent d'évaluer clairement l'état de leurs infirmités. On laisse aux thérapeutes de la parole la double charge du diagnostic et du traitement.

Les classifications des infirmités motrices cérébrales et des troubles de la parole qui ont été trouvées utiles dans les dispensaires d'Edinbourg sont décrites. On s'est efforcé, en les utilisant, de définir et de relever les troubles habituels de la parole observés chez les enfants atteints de différentes formes d'infirmités motrices cérébrales. Les troubles de la parole présentés chez *258* malades vus au dispensaire d'Edinbourg du Scottish Council for the Care of Spastics sont décrits.

Chez les hémiplégiques, il faut considérer le retard du développement de la parole consécutif à un retard mental comme une cause bien plus fréquente de troubles de la parole que la dysarthrie, la dysphasie ou la dysrhythmie. Les enfants atteints d'infirmité motrice cérébrale bilatérale des membres sont plus souvent des dysarthriques que des hémiplégiques,

mais le retard du développement de la parole secondaire au retard mental ou à une audition défectueuse est encore plus important. Un pourcentage élevé de malades atteints d'infirmité motrice cérébrale bilatérale présente des troubles de la parole dus à un certain nombre de causes secondaires. Il est important de tenir compte de ce fait lors du diagnostic et du traitement de la parole chez les infirmes moteurs cérébraux.

ZUSAMMENFASSUNG

Beschreibung und Klassifikation der gewöhnlichen, mit Zerebrallähmung verbundenen, Sprachstörungen.

Die Sprachstörungen bei Kindern mit Zerebrallähmung werden oft von den Ärzten vernachlässigt, wenn sie versuchen, den Zustand der Störungen klar abzuschätzen. Man lässt den Sprachtherapeuten die doppelte Bürde der Diagnose und der Behandlung.

Klassifikationen der Zerebrallähmung und der Sprachdefekte des Kindes, die man in den Edinburger Kliniken nützlich gefunden hat, werden beschrieben. Bei ihrem Gebrauch hat man sich bemüht, die üblichen Sprachdefekte, die man bei Kindern mit verschiedenen Formen von Zerebrallähmung auffindet, zu bestimmen und zu erklären. Sprachstörungen von *258* Patienten der Edinburger Klinik des Scottish Council for tne Care of Spastics werden beschrieben.

Bei Patienten mit Hemiplegie ist Verspätung der Entwicklung der Sprache durch geistige Rückständigkeit eine viel häufigere Ursache der Sprachstörungen als Dysarthria, Dysphasia oder Dysrhytmia. Kinder mit doppelter Zerebrallähmung der Glieder sind öfters dysarthrisch als Kinder mit Hemiplegie aber die Verspätung der Entwicklung der Sprache durch intellektuelle Verspätung oder Hördefekt ist noch häufiger. Ein grosser Teil der Patienten mit bilateraler Zerebrallähmung weisen Sprachstörungen auf, bei deren Entstehung mehrere Ursachen mitwirken. Es ist wichtig, diese Tatsache zu erkennen, wenn man versucht die Sprachanomalien der Patienten mit Zerebrallähmung zu diagnostizieren und zu behandeln.

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L'Hôpital Des Enfants Malades, Paris

In the 18th century children's hospitals were non-existent. There were no treatment centres but only charitable foundations for poor children and orphans. The Hospital for Sick Children in Paris was one of these charitable foundations from 1724, under the name of l'Hôpital de l'Enfant Jesus. Diverted from its original purpose by the Revolution in 1799, it was applied definitely to the treatment of sick children of both sexes in 1802. It was then the only children's hospital in Europe.

In 1820 the hospital consisted of 22 wards with 500 beds. At that time the walls were tiled and special wards for patients with contagious diseases were set up. Only the entrance door and the grills of the Louis XVIth period now survive, but before 1958 the central corridor passed under an ancient pavilion with marble pillars on either side inscribed with the names of house-physicians who had died of diphtheria contracted in their duties. There was also a bas-relief of Dr. Roux, inventor of anti-diphtheria serum.

About 1900, the 800 beds of the hospital were divided into surgical and medical. Patients with contagious diseases occupied relatively modern buildings. Later, the increase in the population brought about a reorganisation of the services and, around 1920, the building of the three-storey Blumenthal Wing for otorhinolaryngology was started.

The ancient buildings have since been modernised. The patients have been isolated in glass cubicles, which only people wearing overalls and masks may enter. Between 1940 and 1945, a huge surgical building of five storeys was constructed with all the services under the direction of one surgical Professor. The laboratories are in the basement; on the ground floor there are the outpatients department for orthopaedic cases and the X-ray department. Three storeys contain the beds for surgical patients, completely isolated by glass partitions, with air-conditioning and an oxygen supply at the foot of each bed. On the fourth floor there are four operating theatres with oval vaults reflecting the light on to the operating tables. Under the vaults is a room where the students can follow the operations by means of microphones and portholes. On the top floor are rooms for archives, secretariat and a library.

Recently, an important new medical department has been erected, including, in two buildings, all the newest features of a modern clinic—total isolation of each child; all the essentials for the care of premature infants and toxic cases; a large amphitheatre for lectures and film projection; and annexes containing laboratories, X-ray department and offices.

The advances achieved in 60 years should not make us forget the Masters of the past, who, without any X-rays or laboratory reports, were able to treat so many children successfully, thanks to their wonderful clinical sense.

Leon Tixier
Honorary Physician to the Hospital

The picture of the Hospital on our cover was drawn by M. Claude Thiberville.

What's to be done?
A real-life problem and some replies

THIS is the second instalment of what we hope will become a practical feature of the question-and-answer type, the first question having arisen spontaneously out of a puzzling case described by Dr. C. Elaine Field (*Cerebral Palsy Bulletin* Vol. 2, 1960, p. 89). Here the puzzle has been set by Dr. Guy Tardieu, of the Hôpital Raymond Poincaré, Paris, and has been circulated to some likely solvers. Further questions suitable for similar treatment will be welcomed.—*Editor.*

The Problem . . .

Danger of dislocation of the hip in asymmetrical spasticity of the thigh adductors.

Dr. GUY TARDIEU *writes:*

This type of case often puts me into a difficulty. The asymmetrical spasticity of the hip adductors provokes:

(1) pelvic obliquity and functional shortening of the limb;

(2) consequently, aggravation of the equinus, which arises partly as a compensation and partly as a result of the spasticity of the soleus and gastrocnemius; and

(3) a danger of progressive subluxation and finally dislocation of the hip.

The various methods I have used hitherto do not seem satisfactory. Stretching of the muscles by physical therapy is difficult, especially if the spasticity appears only on standing, because we have to stabilise the pelvis in the horizontal position.

The Phelps long brace with pelvic band, so useful when the spasticity of the adductors is symmetrical, does not prevent the pelvic obliquity in these cases. According to W. Phelps, the diagonal 'well leg' traction strap solves the problem. Unfortunately, however, it is very uncomfortable for walking, and in one case the subluxation of the hip has been increasing during this treatment, so I did not dare to experiment with it again.

Surgery also presents difficulties and dangers. Of course, if the resistance to abduction is due to contracture from muscle fibrosis one can easily do a myotomy and make the affected muscle or muscles exactly the desired length, but such cases are rare. If the spasticity is of the gamma, purely myotatic, type, present in the lying position, it disappears under anaesthesia, so that the surgeon no longer knows what length to give the muscle. The difficulty is still greater if the spasticity is of the alpha type, appearing chiefly or only on standing. In doing a myotomy, or crushing the anterior branch of the obturator, there is a risk of doing too much or not enough. The bilateral operation is dangerous, because the postural power of the gluteus medius is not easy to test in such cases. There is a great risk that the pelvis will no longer be stabilised. Lowering the insertion of the gluteus medius is theoretically more satisfactory but not always successful.

What is to be done?

. . . and some Replies

1. *From* W. J. W. SHARRARD, F.R.C.S., *Assistant orthopaedic surgeon, Sheffield Royal Infirmary and Children's Hospital.*

The mistake here lies in the assumption that spasticity of the thigh adductors *per se* will result in dislocation of the hip. This is not so. Dislocation of the hip arises in patients in whom there is *postural weakness of the gluteus medius* relative to the strength of the hip adductors, whether the adductors are spastic or not. It is, of course, often found that severe spasticity of the adductors is associated with strong adductor muscles and functionally weak or paralysed gluteal muscles. Valgus deformity of the neck of the femur due to the same gluteal weakness (similar to that seen in other paralytic dislocations, e.g. in meningocele) increases the tendency to dislocation.

In my experience, splintage, especially in a unilateral case, is never satisfactory except as a temporary measure and surgery is always needed eventually.

The aim of surgery is two-fold:

(1) To correct any muscle shortening or bony deformity; and
(2) if possible to balance the adductor/abductor power.

Muscle shortening of the adductors is, as Dr. Tardieu says, easily corrected by tenotomy or myotomy of the adductor muscles in the groin. This in itself will reduce the power of the adductors to some degree and I have never known simple tenotomy alone to result in too much weakening so that the hip goes into abduction. Varus osteotomy of the neck of the femur with nail-plate fixation is also a perfectly safe procedure where there is valgus of the neck of the femur and much reduces the liability to dislocation.

A more difficult problem is that of restitution of balance of muscle power, either in company with the tenotomy or in cases where there is not yet any contracture. It is not easy to assess the power of the gluteal muscles when there is considerable spasticity of the adductor muscles, but instillation of local anaesthetic, first into the anterior branch of the obturator nerve, and then into the posterior branch often makes the assessment of the gluteal power much easier and also allows an assessment to be made of the possible effects of dividing or crushing one or both of these nerves. This also gets over the difficulty described by Dr. Tardieu of spasticity that arises mainly on standing, since a trial of this can be done after the anaesthetic has been put in.

In many cases, the gluteal muscles will show moderate power (3 or 4) such that division of the anterior branch of the obturator nerve alone will be sufficient to restore the balance of power. Only very rarely, if ever, is division of both branches of the obturator nerve indicated, since this would imply complete paralysis in the gluteal muscles and in such a case tendon transplantation is clearly indicated. These latter cases often also have a problem of flexion contracture and spasm of the iliopsoas muscle, and a transplantation of the ilio-psoas muscle to the greater trochanter through a hole in the ilium *posteriorly to the hip* has given a number of very satisfactory functional results as well as preventing dislocation. (This is not the same procedure as a Mustard transplant, in which the muscle is transplanted laterally but left as a flexor.)

Lowering the insertion of the gluteus medius is seldom satisfactory, since, in cases where it is necessary because the gluteus muscle has elongated, the muscle is usually a weak one, and moving it distally to shorten it will only temporarily alleviate the situation.

2. *From* GAVIN C. GORDON, F.R.C.S., *Consultant orthopaedic surgeon, Cumberland and North Westmorland area.*

In 1954, Weissman* described a condition of fixed pelvic obliquity in neonates, presumably due to malposition in utero in the last months of pregnancy, which led to

*Weissman, S. L. (1954) 'Congenital dysplasia of the hip.' *J. Bone & Jt. Surg.*, 36-B, 385.

dislocation of the adducted hip if untreated. A mechanism such as this will bring about dislocation before the acetabular roof is fully developed for the orthograde position —that is, before the child walks. But where the obliquity of the pelvis is due to spasticity, which does not become manifest commonly before the sixth month, the obliquity being a still later development, then the acetabular roof will be developed sufficiently to retain the femoral head, except in extreme cases of adduction deformity or where the acetabulum is shallower than normal.

Where the cerebral palsied child with pelvic obliquity is able to walk, the incidence of dislocation of the hip will be slightly higher than in a normal child, because marginal cases of true congenital dislocation of the hip will show less tendency to spontaneous cure.

When the child is unable to walk dislocation will occur more frequently for the following reasons.

Standing and walking are an essential part of the developing biomechanics of the hip-joint and they lessen anteversion, so increasing the stability of the joint. Standing, and the development of a sense of balance, no matter how, is necessary in cases of cerebral palsy. Standing exercises should be started at the optimum time as estimated in the normal child, and they will indicate at an early stage the likelihood, or otherwise, of pelvic obliquity. If pelvic obliquity develops, stretching of the adductors and passive exercises for the adducted hip are indicated; combined with standing practice. No fixed abduction should be used, but a Frekja pillow is useful to control scissoring when the patient is sitting or lying.

Simple subcutaneous adductor tenotomy would be helpful where severe scissoring persisted, but should not be carried out as a routine procedure.

Under these circumstances dislocation of the hip is extremely unlikely and the risk can be ignored except where the acetabulum shows underdevelopment. Treatment would then be as for ordinary cases of congenital dislocation of the hip, and would include procedures such as a shelf operation and/or osteotomy, provided the child's general condition does not preclude a severe surgical procedure. Subluxation of the hip-joint associated with cerebral palsy is said to be present when the head is not fully contained in the acetabulum, but the hips are stable in these cases and in the vast majority remain so.

When the child fails to stand or is not encouraged to do so, the risk of dislocation and subluxation is greater. The biomechanics of the hip-joint, already prejudiced, become disorganised. If dislocation occurs in these circumstances, then no surgical procedure will give a stable hip. Osteotomy of the Lorenz type is the best procedure in these cases, but this should not be performed unless a clear case can be made out that the child will stand and walk, though if personal hygiene is unsatisfactory owing to the hip deformity, a palliative osteotomy of the hip would be indicated.

With regard to the functional shortening of the leg and equinus deformity of the foot, lengthening of the tendo achilles has given poor results in the cases I have operated on, for relapse occurs, and it has been found better to accept the equinus deformity and be content with some raising of the shoe.

Management rather than operation is the first consideration in treatment.

LETTERS TO THE EDITOR

Is There a Syndrome of Brain-damage in Children?

SIR—Dr. Desmond Pond (*Bulletin* Vol. 2, 1960, p. 303), states that the clinical use of the term 'brain damage' would 'seem justifiable only that thereby attention is drawn to factors within the child that may contribute to the behaviour disorder'. With this few child psychiatrists would disagree. In his concern, however, lest brain damage be regarded as the single determinant of a disturbance in the child he runs the risk of appearing to refuse to accord any significance at all to this factor.

The evidence that neurological injury to the brain of a degree less than gross obliteration of function affects behaviour is strong. From the head injuries of the first world war not only limitation of function but also explosiveness of affect and behaviour were recognised and described in adults by, among others, Hughlings Jackson.[1] The prevalence of epidemic encephalitis in the 'twenties left a residue not only of neurologically damaged and psychiatrically ill adults, but a group of children with gross and severe behaviour disturbance. The description of a rehabilitation programme for these children by Bond and Appel[2] gives a useful picture of these cases. Bender[3] described as prototype the cases deriving from epidemic encephalitis and instanced other virus encephalitides, those associated with pyogenic infection, burn encephalopathy, traumatic states and anoxaemic conditions as being liable to lead to similar disturbances in children. More recently Binns[4] has described eleven cases of acute encephalitis with sequelae.

The concept of there being sufficient similarity between the effects of the brain damage in epidemic encephalitis and other forms of encephalitis and encephalopathy to allow of generalisations as to the effects of this factor derives from Bender. She states in addition, however, that 'the final career of the individual is as much dependent on personality configuration, constitutional endowment, infantile personality factors, family and social relationships—as in organic damage, except when that is severe.'[3]

The general agreement in the truth of this latter statement is not, however, incompatible with the direct effect of injury to the brain.

In respect of the psychological evidence, particularly in relation to perceptual anomalies, this is much more strongly supported than Dr. Pond implies. In common with the EEG, however, the psychological test seldom gives definite information when historical, clinical and neurological evidence is dubious. Psychological testing in this field is, however, extremely important in that special education techniques may be indicated in the rehabilitation of the child.

It is, I think, significant that the conditions instanced by Bender as liable to be associated with subsequent disturbances are those in which diffuse or scattered minor damage to the brain might be assumed. This would render it more probable that particular areas or tracts would be involved whatever the causes of the scattered damage. I agree that general or focal maldevelopment is seldom associated with the type of disturbance indicated.

I regard it as clinically useful to relate together, in the motor field, tics and hyperac-

1. Selected Writings of John Hughlings Jackson. Ed. by James Taylor. *London:* Hodder and Stoughton, 1932.

2. The Treatment of Behaviour Disorders Following Encephalitis. *New York;* Commonwealth Fund, 1931.

3. Bender, L. 'Organic brain conditions producing behaviour disturbance.' *In:* Modern Trends in Child Psychiatry. Edited by Lewis and Pacilla.

4. Binns, R. Report of the Adelaide Children's Hospital for 1954, Vol. 2, p. 172.

tivity; in the intellectual field, limitation of intelligence or perceptual anomalies; in the affective field, lability of mood and in the personality loss of acquired inhibitions and controls with neurological or other evidence of brain damage, and I regard the last as a causative factor.

That any one of these—in particular hyperactivity—may be due to other causes is undoubtedly true.

That all child psychiatry must involve a global approach is undoubted. That brain-damaged children are 'more global' than undamaged ones is perhaps true.

I have discussed this with Dr. Pond from time to time. Perhaps, Sir, I might through the courtesy of your columns, now let him 'have it in writing'.—Yours, etc.

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SIR—Dr. Desmond Pond poses a question which, if stated in more general terms, has an application to all diagnostic labels of psychological problems of childhood. Such labels often over-simplify the problems involved beyond the point of clinical usefulness where the individual is concerned, and may pave the way for misunderstandings to become established.

The difficulty with the clinical term 'brain-damage' as a name for a syndrome is that it is used as an all-inclusive diagnostic label representing both a group of symptoms and a dynamic concept. Unfortunately even this usage has not been consistently maintained, resulting in confusion when the term is sometimes used in the narrower sense as the single cause of a group of symptoms. Furthermore, this narrow usage has sometimes engendered the unhappy connotation that any child with such a diagnosis is unable to benefit from psychotherapy.

The existence of actual brain-damage does not in itself exclude a psychological problem. On the contrary, the resultant capacities for brain function may, and often do, present the child with a complex handicap of varying degrees with which he must deal. Moreover, the damaged brain of the child does not remain static. While the original injury to the brain may have been of limited intensity duration and extent, its importance lies in its subsequent effects on the child's psychological maturation and development.

For example, the timing of the brain injury with respect to the stage of development of the child may well be crucial. An injury which may be trivial at, say, 6 years of age may have a devastating effect on an infant aged 6 months, at least from the point of view of subsequent development. Yet here too caution is necessary, since as maturation proceeds new capacities may emerge which will enable the child to compensate or overlay an earlier injury.

The significance of this dynamic time factor lies in the different meanings the original injury and any consequent handicap had or continues to have for the child as well as his parents.

Brain damage itself may or may not manifest itself as a neurological deficit. Some children with presumed, or initial, or later persistent deficit may also exhibit certain common behaviour traits. It is this whole complex that constitutes the clinical syndrome under discussion.

It should be emphasised that this syndrome is not caused by the single onslaught of an injury to the brain. To begin with, our diagnostic skill is often unable to detect, or at least differentiate, minimal brain-damage in what is after all a complex functional problem. Then, as Dr. Pond points out, there is no evidence at present to prove a direct correlation between a behaviour item and a manifest neurological deficit, let alone an apparently recovered neurological deficit or an earlier presumed deficit. But here again lies the danger

of falling into the trap of supposing a simple static and direct relationship as the only valid operational basis, a legacy from an earlier period in medicine.

In the case of the so-called brain-damage syndrome, one can say that in a significant number of cases both an injury to the brain of varying detectable degrees and a group of behaviour characteristics are both present. From this observation one may suggest the hypothesis that in some cases this injury may manifest itself, under certain conditions, with a peculiar group of behaviour characteristics. We may add that this hypothesis can be a profitable operational basis from which to work until more refined techniques of examination and methods of research become available.

The syndrome of brain-damage can now usefully be regarded as a result of a dynamic interaction involving numerous factors, such as the degree and timing of the original brain-injury; the psychological state of the child and his parents before, during and after the injury; the meaning of the injury to both child and parents; the prevailing conditions which may aggravate or relieve the symptomatic behaviour; and the attitudes adopted by both child and parents in attempting to deal with the situation.

It is indeed a current problem in child psychiatry that diagnostic classifications of a mixed nature are used to embrace a wide range of psychological problems. For this reason alone the usefulness of any diagnostic term as applied to a particular child may be seriously weakened, as well as bringing with it the further danger of treating the diagnostic label instead of the child.

To answer Dr. Pond's question, there *is* a syndrome of 'brain-damage' in children, but it is not a simple concept. The aetiological factors are both physical and psychological, influenced by both endowment and environment, changing with maturation and development. To merit usefulness the name of this syndrome should connote this broader dynamic concept. Perhaps the present name might be modified in such a way as to reflect this concept —e.g., 'Brain injury-Reaction Syndrome.' Incidentally, one added advantage to such a concept is that it permits greater flexibility for increased therapeutic efforts.—Yours, etc.

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SIR—Dr. Desmond Pond asks rhetorically 'Is there a Syndrome of Brain Damage in Children?' and makes his own position very plain with the comment, 'It is time this idea of a syndrome was scotched before it gets too firmly entrenched in the literature.' As one of the sinners who has recently imputed to these children the very characteristics which he lists—intense anxiety, 'catastrophic reactions', distractability, &c.—I feel obliged to reply to his criticisms. But first let us be clear that we avoid semantic confusion.

I agree fully that there is no evidence to suggest the existence of *a syndrome* of brain damage. But I do not agree that the behaviour disorders of brain damaged children 'are largely the result of the handling or mishandling . . . by parents and society generally'. My criticism of the evaluation of these cases in child-guidance clinics is not that the brain damage is held responsible for symptoms which are really psychogenic; it is that too often the presence of a neurological lesion is not even suspected, and when recognised it is not given adequate weight in the total case assessment. As I indicated in my recent paper[1] there has been very little systematic study of the influence of brain damage on personality development, and I suggested that certain of these children show a form of impaired ego control which causes them *to react to stress in an extreme and characteristic fashion.* In other cases where there are gross perceptual defects this, I believe, has a direct bearing on subsequent distortions in personality development—e.g., as a result of misinterpretation

1. 'Psychodynamics of Brain-Damaged Children: A Preliminary Report.' *J. Child Psychol. Psychiat.*, 1960, 1, 203.

of reality experiences from infancy onwards. Both these types of effect, quite apart from learning disabilities, may be aggravated by environmental factors but are not caused by them, and it is distressing to witness in our clinics the misplaced efforts at modifying parental attitudes in the mistaken belief that it is there that the pathogenic influences lie.

I feel very sorry for many parents faced with these complex problems of child-rearing and try whenever possible to help them by explaining the nature of the child's disability, and supporting them in providing for that child's special needs. It is, of course, correct that sometimes the child is brain-damaged, and in addition there is a grossly disturbed parent-child relationship, not just the reaction to having a handicapped child. Even here, I question how justifiable it is to undertake major case-work or psychotherapeutic procedures with parents without a realistic appraisal of the child's prognosis. This means that the parent-therapist must have a fair knowledge of paediatric neurology.

While hesitating to challenge Dr. Pond on a purely neurological issue, I nevertheless question whether clinical neurology is sufficiently developed, especially in the examination of very young children, to demand as a first criterion of brain-damage 'unequivocal neurological signs'?—yours &c.

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Autofaradisation for Paretic Muscles

SIR—In rehabilitating patients with paralytic disorders, such as poliomyelitis, the major problem is which of the available forms of therapy to use for initiating the recovery of function, particularly when little or no clinical recovery in muscle function can be detected. In addition to the more usual forms of exercise, electrical stimulation can be used to initiate contraction of viable muscle, thus assisting re-education of the voluntary use of the muscle by the patient.

In present methods of electrical therapy, the intensity, duration and rate of repetition of the stimulus have received more attention than the rhythm in which these impulses are applied. It seems likely that motor units normally discharge rhythmically, the rate probably lying within a definite frequency. A method known as 'autofaradism' has been developed to imitate this rhythmic stimulus. Electrical potentials from a normal contracting muscle, preferably a corresponding one on the other side of the body, are used to stimulate the paretic muscle. These potentials are detected by electromyography; the electrical impulse travelling to a muscle is located with electrodes, amplified, and recorded on magnetic tape, since it is inconvenient to stimulate directly from the individual muscles. This recording is preserved and when required is fed into an electronic translator which converts the recorded potentials simultaneously into faradic current and sound played through a loudspeaker.

Therapeutically, the patient listens to the record of the muscle alternately contracting and relaxing on the loudspeaker, and simultaneously feels the faradic stimulation of his own weak muscle. At the same time, a physiotherapist moves the joint involved through a full range of passive movement. Where unilateral lesions are being treated, the corresponding muscle on the normal side is contracted at the same time. This combination of hearing, feeling and active and passive movement all act together to facilitate the maximum possible response from the paretic muscle. Each treatment lasts 2–5 minutes and is repeated twice daily for several weeks or until voluntary activity returns.

Considerable improvement in muscle power is said to be obtained by this method, even after several years. Sufficient power to eliminate the effect of gravity was recovered in 6

out of 7 patients treated, in whom no clinical volitional power had previously been detectable.

No controlled trial has yet been attempted, but this new method is described here because it suggests possibilities for accelerating the recovery of motor function in partially paralysed muscles.—Yours, etc.

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Growth Control of the Sympathetic System by a Specific Protein Factor

SIR—Earlier studies in chick embryos demonstrated that it is possible to evoke overgrowth of sympathetic ganglia by chemical substances isolated from different biological sources (mouse sarcomas, snake venom, and mouse salivary glands). In all cases the active agent promoting the overgrowth was identified in a protein fraction.

The investigations were then extended to mammals, and it has been shown that a similar effect on the sympathetic ganglia is produced in newborn and adult mice by daily injections of the same nerve-growth agent. The experiments in postnatal life were performed with the protein fraction isolated from mouse salivary glands. The injection of this agent causes a 4- to 6-fold increase in the size of the animals' sympathetic ganglia and a great increase (hyperneurotisation) in the nerve-supply of their viscera. Other components of the mice's nervous system are apparently unaffected. The injected animals are as healthy and vigorous as untreated controls.

An antiserum to the protein isolated from mouse salivary glands has been prepared by Dr. S. Cohen, and the injection of a small amount of this into newborn mice and other mammals results in the near-total destruction of the sympathetic ganglia. The sympathetic system of adult mice is also severely affected by this treatment. The injected animals do not differ from controls in any other respect.

Pharmacological and electrophysiological experiments on mammals experimentally deprived of their sympathetic nervous system are now in progress.

Those interested in this work will find more details in the paper I read at the First International Conference on Congenital Malformations in London on July 20, 1960, which will be appearing in the Proceedings. I have also attached a brief bibliography.—Yours, etc.

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BOOK REVIEWS

Physical Disability—a Psychological Approach

By BEATRICE A. WRIGHT.

New York: Harper & Brothers, 1960, pp. 408, S6.

Reviewed by Mrs. E. M. MASON.

Dr. Wright here sets out to show how the psychological reaction and underlying attitude towards physical disability, in both the disabled and the general public, affects rehabilitation and education. To English readers the use of sociological jargon may seem a little tiresome, but Dr. Wright's ideas should prove both stimulating and helpful to all those concerned with the upbringing, education or rehabilitation of the physically disabled, and extracts from autobiographies and records bring life to her thesis.

All societies, in Dr. Wright's view, over-emphasise the importance of normal physique, and this creates unnecessary difficulties for the disabled person, who is under pressure to act as if he were physically normal. This in turn leads to conflicts which may result in attempts to deny or conceal the disability, or to an aggressive and over-competitive attitude. The suggestion that this emphasis on the normal also affects the upbringing and education of the disabled is of considerable importance. In giving so much time to the training of blind, deaf or crippled children and adults in 'the normal techniques of living', we may well miss the real purpose of education—the development of clear thinking, the communication of ideas, and the ability to get on with others. Instead of focusing on the teaching of lip-reading to the deaf or on walking to the crippled, Dr. Wright suggests that we should find the method of communication or of motility which comes most easily to the individual. She gives examples of people who discard techniques learnt so laboriously in favour of their own special way of overcoming their disability. In working with blind children one cannot but be aware of how greatly the sighted world's standards of behaviour, vocabulary and concepts are imposed on and accepted by them.

Taking the abilities and disabilities of each individual as our guide in education and rehabilitation, rather than the standards of normal development, Dr. Wright suggests that the disabled person can be helped to change his values so that physique counts for less in his concept of himself and his other abilities are included. She gives interesting examples of the way in which physically disabled children think of themselves as being like their companions; in spite of all the evidence to the contrary they do not look on themselves as deformed, and they need to be helped by their parents to understand and accept their physical disabilities. She describes the ease with which a small girl in a nursery school, who has had this help at home, responds to curiosity about her braces by saying in a matter of fact way that she has cerebral palsy, and then continues with her play. Illustrations are also given of children who have been encouraged in their denial of their disability by over-protection at home, and who are suddenly and cruelly made aware of it by other children when they go to school. They have then, painfully, to come to a re-assessment of themselves and their abilities and disabilities.

In discussing programmes of rehabilitation and education, Dr. Wright emphasises a phenomenon not always sufficiently recognised—that a period of mourning, depression or withdrawal frequently follows the realisation of being physically disabled, because this is felt as a loss of bereavement. In some people this period of mourning is a necessary phase of healing which must be experienced before re-education can begin.

Dr. Wright again stresses the 'idolising' of normal physique when she discusses the social attitude to disability. This tendency, she suggests, results in a negative reaction to any physical deviation. She finds that people may react with aversion, teasing or bullying, or by an over-pitying or over-positive attitude, equally unacceptable to the disabled. The tendency to a 'spread of disability' can result from these attitudes; we have probably all witnessed situations such as those cited by Dr. Wright in which people talk in front of the blind as if they were not present or could not hear; treat the deaf as if they were stupid, or crippled people as if they were totally handicapped in life. This attitude can easily influence the disabled person, so that in planning his career, or in his social life or in contemplating marriage, he over-estimates the effect of his disability.

In considering the possible causations of the over-estimation of the normal in physique, Dr. Wright briefly touches on the question of unconscious motivations. It is certainly difficult to account for the irrational anxiety, aversion or even horror which the sight or experience of some bodily defects can arouse—particularly in childhood—unless we pre-suppose deep and archaic fears, of which we are unconscious, but which can be touched off by the sight of the loss of some part of the body. Although she puts forward this theory—that the source of the strong emotional reactions to physical disability is in the unconscious mind and is therefore not recognised by the disabled or by society—Dr. Wright does not follow this up in her recommendations for educating children in helpful attitudes towards the disabled, which remain therefore rather superficial.

In her chapter on *The Parent as Key Participant*, Dr. Wright makes some excellent practical suggestions on how parents can help their children, but again she does not allow sufficiently for the deeper problems which lie behind the attitudes she discusses, such as over-protection and over-insistence on independence in bringing up children with physical disabilities.

This book makes a valuable contribution in drawing attention to the need for considering the psychological reactions to physical disability and for asking ourselves whether we do not sometimes go too far in applying the standards of the physically normal to the education of the physically disabled. It also paves the way to a further consideration of possible methods of dealing with irrational and harmful reactions to disability, both in the individual and in society. E. M. MASON

Health in Childhood

By PROF. RICHARD W. B. ELLIS.

London: Pelican Books, 1960, pp. 251, 5*s*.

The foreword to this book, which is aimed primarily at the 'man in the street', might give the impression that it deals with the common problems and disorders of childhood arising in a family setting. Professor Ellis has allowed himself to range far beyond the confines of childhood health and disease as they might present themselves to an individual parent.

In eight chapters he sets out some of the current views on the biological aspects of childhood, the genetic make-up of the individual, world problems of nutrition, changing patterns of growth and develop-

ment, and some of the difficulties of the handicapped child.

There is only one chapter, on *Health and Disease*, that is at all 'Spockian' in its compass. Here the current paediatric views on the tonsil and adenoid problem, enuresis, rheumatism and some other common disorders are put forward in a restrained and easily understood manner.

For the rest Professor Ellis reminds us of the world problems in child health with references to neonatal tetanus, malaria and kwashiorkor. Our enlightened views on maintaining the nutrition of young people by rationing and priorities are quite recent history, as witnessed by the fact that during the siege of Paris in 1870 hardly a child under three survived.

When discussing education Professor Ellis suggests that there is a tendency to start formal schooling before many children are really ready for it—a view held by some 'educationists' but seldom expressed. He regrets the passing of the apprentice situation in the home, where the children of both sexes learned from their parents the basics of a trade on the one hand and of domestic skills on the other. The practical difficulties of providing part-time education and part-time employment, together with the significant advancement of the age of reaching maturity, may well be retarding the adolescent's social development, thus leading to the present problems at this age.

Perhaps the chapter on *The Hazard of Birth* is a trifle too obstetrical—one wonders what the man in the street is to learn from the indications for forceps delivery, but this is soon followed by a disarmingly simple account of the physiological changes experienced by the foetus at birth, which makes amends.

In broader fields we learn how primitive polygamy gave place to a more civilised monogamy and has now been followed by 'successive monogamy'; this and other factors have led to the deflation of the father-figure—an appalling thought! Parents of boarding-school children who are not gaining weight are taught to ask three questions: Has the school diet been adequate? Has the school been over-exercised? Have the children been getting enough sleep?

The final chapter, on *The Handicapped Child*, briefly displays the problem as a whole and lays suitable emphasis on emotional maladjustment. Two quotations from this chapter might encourage *Cerebral Palsy Bulletin* subscribers to buy this excellent book.

'Since the policy of a minimum wage is ultimately liable to prejudice the employment of the physically or mentally defective, the fate of the handicapped child who has received special schooling to the age of sixteen is one which the Trades Unions should have constantly under review.'

'The happily married, stable, devoted mother of no outstanding intellectual attainment . . . is more likely to raise stable children than one who is emotionally frigid or over-anxious, however profound her reading in child psychology.'

B. S. B. WOOD

Mirage of Health

By RENÉ DUBOS. *London:* George Allen & Unwin, 1960, pp. 221, 21*s*.

THIS is a remarkable book, and for many reasons. The range of allusion and breadth of reference—from Lao-tzu and the Yellow Emperor to Ibsen and Dostoevsky, from Propertius and Juvenal to Katherine Mansfield and D. H. Lawrence—make one wonder how a single man could at the same time be so widely read and be one of the world's leading microbiologists. The easy style belies the author's French origin; indeed, many scientists whose mother tongue is English would learn a great deal from his clarity and readability. Perhaps the most remarkable aspect of the book, however, is its quiet optimism.

Dubos stands back and looks at Man as an animal and examines Man's past and future against the background of his own

society and the rest of evolving life on earth; yet, unlike the Huxleys, Orwell, Wells and other authors who have written similarly, he does not overwhelm the reader with a sense of gloom and futility. Dubos' thesis is that although every creed and cult has had in lore or mythology its imaginary golden age when all was peace, health, prosperity and happiness, such 'Gardens of Eden' have seldom existed, nor are they likely to exist because Man's restlessness and creative curiosity are continually changing his environment in totally unforeseeable ways, and as one disease is conquered another appears in its place.

'The introduction of inexpensive cotton undergarments easy to launder, and of transparent glass that brought light into the most humble dwelling, contributed more to the control of infection than did all drugs and medical practices.' Yet the Industrial Revolution which made these materials available brought also atmospheric pollution, which Dubos identifies with the apocalyptic horseman invoked by St. John the Divine's Fifth Angel: 'There arose . . . the smoke of a great furnace and the sun and the air were darkened by reason of the smoke.'

Again, the importation of the American potato to Europe in the sixteenth century, followed by the potato blight, contributed to the Great Famine of 1846, and the consequent immigration of millions of Irish back across the Atlantic led to the great epidemic of tuberculosis in the Eastern United States in the early 1850's. Unforeseen consequences will also result from the change Man is enforcing on his own genetic make-up, because by drastically reducing infant mortality he is for the first time breeding a race which does not have to withstand the rigours of natural selection, a race which is biologically unfit.

Dubos is careful to distinguish between health and lack of disease. 'Too often,' he writes, 'the goal of the planners is a universal grey state of health corresponding to absence of disease rather than to a positive attribute conducive to joyful and creative living. This kind of health will not rule out and may even generate another form of ill, the boredom which is the penalty of a formula of life where nothing is left unforeseen.'

The book is not entirely without blemish; the author is inclined to repeat himself as well as being guilty of minor inaccuracies. It is untrue to say of the era when Keats wrote his *Ode to Autumn* that 'to poetic souls of the period autumn did not signify the time of crops and abundance but the death of everything in nature.' The Royal College of Physicians' building in Pall Mall is referred to as the Academy of Physicians, and the great London smog of December 5–10, 1952 is placed in 'late December'. Typographical errors and misprints are also far too numerous for a book which is otherwise well produced. These, however, will doubtless be put right in the many subsequent editions to which it will surely run, for it combines educative value and pleasure and should be read by doctor, layman, student and any other intelligent person who feels his faith in Man is failing.

ROY M. ACHESON

Psychological Problems in Mental Health

By SEYMOUR B. SARASON with THOMAS GLADWIN. *New York:* Harper & Brothers, 1959, 3rd ed., pp. 678, £2 12*s*.

This edition differs from the preceding one in having a monograph entitled 'Psychological and Cultural Problems in Mental Subnormality' tacked on to the end. The author apologises nicely for not having combined the previous and more recent work into one: '. . . neither Dr. Gladwin nor I could generate the slightest bit of enthusiasm for the job of writing an integrated text'. What a pity!

In the first part of the book much of the recent psychological literature of mental deficiency is briefly reviewed. The proper use of a large number of psychological

tests and their interpretation are discussed. In addition, many problems are aired which are of interest to others, besides psychologists, who work with mentally defective patients. For example, the first two chapters are concerned with the criteria of mental deficiency and its classification in various ways. There are chapters on 'The Interpretation of Mental Deficiency to Parents' and 'The Problems of Institutionalisation', which might be considered to be primarily social aspects, and on 'Cerebral Palsy' and 'Problems in the Diagnosis of Birth Injury', which might be considered predominantly of medical interest.

In the second part of the book many of the subjects discussed in the first part are rediscussed, though with new facts and figures and with greater incisiveness. Thus 'The Criteria of Mental Defect', 'Heredity and Environment', and 'Cultural Background Factors Affecting Performance' are some of the headings. The various aspects of heredity and environment in relation to intellect, social adequacy and the problems of obtaining objective methods of measuring these functions are fully discussed. For example, there is a small highly informative section on various aspects of bilingualism.

The main use of the volume in its present dichotomy will be as a reference book. Neither part of it is well written and the first part is discursive and would improve by being more closely organised. When dealing with non-psychological topics it tends to be somewhat naïve. It does review a great deal of literature, however, and the two bibliographies will be found of value even though that belonging to the first part is already out of date. It is to be hoped that the authors will be able to 'generate some enthusiasm' before the next edition.

T. T. S. INGRAM

Understanding and Teaching the Dependent Retarded Child

By LOUIS E. ROSENZWEIG and JULIA LONG. *Darien, Connecticut:* Educational Publishing Corporation, 1960, pp. 224, 30*s*.

This book is intended as a practical guide for parents and teachers in the training of the so-called trainable but not educable mentally handicapped child. The authors divide people with I.Q. 0–75 into four groups; the Independent of I.Q. roughly 65–75; the Semi-dependent (I.Q. 50–65); the Dependent (I.Q. 30–50); and the Custodial (I.Q. 0–30). The first two groups the authors feel often achieve an independent livelihood. The last group requires mainly nursing care. It is the training of the third group, the Dependent Retarded Child, which is the subject of this book. Training in this group should aim at a mental and social level of 6 to 8 years of age. The training skills are divided into Self-help, Social, Motor, Academic and Vocational. The learning of simple skills, such as feeding and dressing, are mentioned, as well as more complicated achievements such as using the telephone and understanding bus fares. There are suggestions for leisure activity and holidays. Numerous references and a carefully described list of equipment are included.

In England this group of children are outside the provision of the educational system and are the responsibility of the mental health authorities. These children attend occupation centres or schools provided by mental deficiency hospitals, and they are trained by people who have mostly had only a shortened period of training or are untrained.

There is a dearth of books of this type, and all teachers would be well advised to read this book and study the goals of training. It will encourage them to feel that their jobs are now the subject of skilled thought and research. They can work out whether children in their centres could reach these goals with the given oppor-

tunity. A criticism that could be raised is the emphasis on teaching. We are now becoming very interested in the training of mentally retarded children through free play and experimentation. It is refreshing to find two writers of high academic standard giving thought and time to the serious but neglected problem of the training of potentially useful citizens; and producing such a practical and readable book.

GRACE E. WOODS

Baby Talk

By MORRIS VAL JONES, PH.D.

Springfield, Illinois: Thomas, 1960, pp. 93, 36*s*.

The author of this book is at present the specialist in speech and hearing to the school for cerebral palsied children in San Francisco, but this book is not concerned with speech therapy for cerebral palsy, nor does it discuss developmental aspects of speech acquisition in early childhood. As Dr. W. J. Wedell indicates in his preface, it is intended 'as a guide for parents who want to help their child to overcome non-organic articulatory errors'. He considers that any child who cannot make himself understood to members of his own family by the age of 3 years should consult a speech therapist. This book, however, is directed towards the treatment of children of 5 years and over who show persistent immaturities of articulation but are otherwise normal. Within these narrow terms of reference it succeeds admirably. It offers authoritative and practical advice which any intelligent parent can appreciate and follow. The British reader may find the numerous analytical 'quizzes' somewhat laboured, and most of the rhymes and stories suggested for speech practice are American. Nevertheless the 4 main 'solutions' offered are universally applicable, since they are founded in good common sense: seek professional help; increase speech stimulation; eliminate other causes, physical, emotional or environmental; and 'do it yourself'. This final section explains simply and clearly suitable methods and materials for the parent to use at home.

MARY D. SHERIDAN

An Introduction to Embryology

By B. I. BALINSKY

Philadelphia and London: Saunders, 1960, pp. 562, 54*s*.

Professor Balinsky does not clearly define the audience for which this new book is intended. Presumably it is primarily for students of zoology, despite allusions to medical students in the preface. Unfortunately, the book lacks the morphological detail of development customarily regarded as necessary in medical training. Moreover, developmental processes are illustrated by the whole range of vertebrates, with frequent references to arrangements in invertebrates. Thus, the book is not suitable for conventional medical courses of anatomy, and it is too large for the usual premedical zoological course.

These remarks are recorded with regret, for the book is a most illuminating introduction to embryology—wide in scope, generously illustrated, excellently documented (21 pages of references) and pleasantly readable. Unlike many texts—and particularly those suitable for medical students—morphological description is throughout associated with physiological processes. Experimental, genetic, hormonal and biochemical data are not briefly

dismissed in chapters rigidly separated from descriptive detail; all is intermingled to give a most lively, dynamic picture of development.

In medical and other teaching apparently irreconcilable schools of thought exist which respectively emphasise factual information and 'principles'. Neither extreme is useful, or indeed possible, without the other. The problem in all teaching is to set the best balance. In this regard, Professor Balinsky's book is excellent. Many of those who teach embryology to medical students would gladly jettison much specific detail to make room for more knowledge of developmental processes. Much morphological detail could be relegated to postgraduate courses, with their specialist needs. Acquiring the intimate knowledge of the development of various systems and organs customarily expected of medical students is a formidable and often tedious undertaking. Exchanging much of this detail (little of which is retained or utilised) for a deeper appreciation of embryological mechanisms renders the task far more interesting and profitable.

The great advances now being accomplished in the genetic and biochemical aspects of development will inevitably alter the content of embryology courses, in medicine as elsewhere. Yet until examination requirements in medicine, and therefore their syllabuses, turn away from the minutiae of structure and towards a more truly biological outlook, texts such as this, excellent as it is, cannot be recommended to medical students as a workbook. Their instructors, however, must surely welcome it. ROGER WARWICK

Pathology of Infancy and Childhood

By AGNES R. MACGREGOR

Edinburgh and London: Livingstone, 1960, pp. 631, 75*s*.

In her preface Dr. Macgregor points out that general pathologists, whose main work and interests are with older patients, are often comparatively unfamiliar with the special aspects of disease in children and particularly in young infants. Her purpose is to provide an account of the morbid anatomy and histopathology of disease in infancy and childhood in a volume of moderate size.

The book is beautifully produced and contains much valuable information. It is eminently readable for pathologists or clinicians, but is not a complete work of reference. This defect is to some extent remedied by the provision of selected references, many of recent date. One of the features of the book are the numerous illustrations and photomicrographs, all of a very high quality.

Four pages are devoted to cerebral palsy but the reader is referred to specialised works for detailed information. Dr. Macgregor draws attention to the present lack of adequate post-mortem information on the disorders which make up this group. Many patients at present die at home, or in institutions without pathological facilities, and this must be remedied if maximum progress is to be made in sorting out the aetiological factors in cerebral palsy.

N. R. ROWELL

Congenital Deformities

By GAVIN C. GORDON

Edinburgh and London: Livingstone, 1960, pp. 136, 37*s*. 6*d*.

This is an unusual book, and one should not be misled by its title into expecting a synopsis of the vast field of congenital deformities or an essay on the basic principles underlying them. It is only fair to prospective readers to explain that the chief theme of the book is congenital dislocation of the hip, and it is in this sphere that the author has most to contribute both as regards original work and

constructive ideas on aetiology.

He analyses the role of acetabular development at the time when the hip-joint opens at about the ninth week of embryonic life, and his study of intra-uterine limb movements in relation to the protective function of the ligament of Bigelow is of special interest.

There are two rather incongruous appendices. In the first, which deals with problems associated with defective blood-supply of the head and neck of the femur, the connection with the main theme is indicated but seems forced. In the second, 'the nature of spasticity and mental processes' is dealt with in six pages. This ambitious project starts with a fascinating train of thought suggested by the finding of normal leg movements in an anencephalic infant, but we are soon lost in a welter of confused discussion on Man's relation to his environment and the profound and enlightening conclusion that 'present views on philosophy, abstract thought and metaphysics will require revision'.

Really Mr. Gordon! But thanks all the same for some stimulating ideas.

R. B. ZACHARY

INTERNATIONAL CATALOGUE OF WORLD MENTAL HEALTH FILMS

A WELL-PRODUCED volume,* sponsored by Smith, Kline and French Laboratories Ltd., and edited by Dr. T. L. Pilkington, contains much valuable information on films relevant to the very broad field of mental health (and illness). Its future users will be grateful for the assiduity of the editor, and perhaps the availability of the catalogue will induce a wider interest in and a greater appreciation of films.

Two practical comments might be made: when editions of catalogues are infrequent sturdy bindings are specially desirable; and this catalogue is larger than most bookshelves and could with advantage be made smaller. Apart from these minor defects, only praise can be afforded the new edition.

Jonathan Gould

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Abstracts

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Spasticity and Rigidity: an Experimental Study and Review

G. RUSHWORTH. *Journal of Neurology, Neurosurgery and Psychiatry*, May 1960, **23**, 99–118.

The definition of the term 'tonus' and the importance of the discovery of the stretch reflex are discussed. The author then reviews the work of others who tried section of various dorsal roots and procaine nerve block to lessen increased muscle tone; this work led to the discovery that the gamma motor fibres to the muscle spindles are the pathways on the integrity of which the stretch reflex depends.

The findings are reported in 50 patients with various types of spasticity, rigidity, or dystonia who had been examined at the Neurological Research Unit, Churchill Hospital, Oxford, since 1954. In most cases a spastic lower limb was tested before and after a block of the motor nerve endings with 1 per cent procaine. Dynamometric and electromyographic readings were recorded. Only one patient showed residual spasticity after the procaine block; it was thought that this case was unusual in that the alpha motor neurones were the pathway and that a type of spasticity had developed which was not abolished by dorsal-root section. In another patient there were decreased spasticity, decreased reflexes, and decreased voluntary power, but all the remaining patients showed abolition of spasticity and rigidity, loss of tendon reflexes, and increased voluntary power.

It is suggested that the significance of these results lies in the preferential blocking of the small gamma motor fibres. This leads to desensitisation of the stretch receptors and so releases motor neurone pathways for voluntary activity which had previously been blocked by stretch reflexes competing for the common route. It is also suggested that the prolongation of the effect of the injection beyond that due to the procaine might be due to the 0.2 per cent chlorocresol in the solution acting like a phenol block.

Janet Q. Ballantine

Note: The newer physiology and anatomy considered in Rushworth's excellent work is fundamental to an understanding of the basic facts of muscle tone. Studies of this nature at the spinal and peripheral levels will surely lead to a much better appreciation of the multiple influences in spasticity, and, we hope, enable an improving approach through medication and physical therapies. We are all hopeful that the Neurological Research Unit at Oxford will be a focal point for increasing enlightenment in neuromuscular physiology and pathology.—*A. V. Neale.*

Reflex Therapy in Spastic Paralysis

Z. MIŘATSKÝ, K. OBRDA, and O. STARÝ. *Časopis lékařů českých*, April 1960, **99**, 520–527.

At the Charles University Neurological Clinic, Prague, the establishment of conditioned motor reflexes has been used in the rehabilitation of motor disturbances in patients with spastic paralysis. An abnormal response in such reflexes (as in the Babinski reflex which is present in these patients) was used, together with normal flexion, tendon, and postural reflexes as unconditioned reflexes. In the course of treatment these reflexes were conditioned to optic and verbal stimuli, their successful establishment being judged by electromyography. By these means it was found possible to improve the mobility

of a spastic lower extremity in 100 patients, using conditioned flexor reflexes, while in 60 patients the mobility of a spastic upper extremity was improved by the use of conditioned tendon and periosteal reflexes. It is suggested that other disturbances of motor function may be successfully treated on the basis of such motor reflexes as can be elicited, including those usually considered to be pathological

G. Hilton

Note: A great deal of 'conditioning' takes place in most forms of treatment in cerebral palsy. Some of the reasons for improvement may be found in the experimental work of the Prague workers outlined here.—*A. V. Neale.*

Use of Phenol in Relief of Spasticity

L. A. LIVERSEDGE and R. M. MAHER. *British Medical Journal*, July 2, 1960, **ii**, 31–33.

The authors of this paper from Manchester Royal Infirmary and Birch Hill Hospital, Rochdale, describe their method of intrathecal injection of 5 per cent phenol in glycerin for the relief of painful flexor spasms. The major hazard is the disturbance of the urinary bladder and its sphincter action and it was found that if the phenol reached the spinal cord itself (at the level of L 1) or descended to the sacral roots urinary retention was likely to occur. The best and safest site for the injection was found to be L 2/3 or L 3/4. The patients were positioned so that the mid-lumbar area was lowest and 0.2 ml. of the 5 per cent solution of phenol was then injected slowly, followed by 0.8 ml. in 30 seconds; altogether 75 injections were given to 32 patients.

The greatest relief was obtained by immobile patients with painful flexor and/or extensor spasms. The condition of such patients was often converted from a painful life in bed to one of moderate comfort in a chair. Conversion from immobility to complete mobility was, unfortunately, rare. The duration of benefit was uncertain and varied considerably from patient to patient, but even if the flexor spasms recurred the associated pain was often reduced. The authors conclude that appreciable benefit can be expected for a period of 6 to 12 months.

N. S. Alcock

Note: A procedure of the kind described here is unlikely to have wide applications in childhood, but it is interesting to note the careful and courageous approach in trying to improve the lot of the unfortunate patient who is gravely troubled by unpredictable or almost continuous painful flexor spasms. In some cases, at least, a grateful relief is obtained. —*A. V. Neale.*

Decrease in Muscle Spasm Produced by Ultrasound, Hot Packs, and Infra-red Radiation

F. P. FOUNTAIN, J. W. GERSTEN, and O. SENGIR. *Archives of Physical Medicine and Rehabilitation*, July, 1960, **41**, 293–298.

Static-force balance was used to measure the resistance to passive lateral flexion of the neck in 17 patients with neck-muscle spasm and to passive extension of the leg in 7 patients with poliomyelitis who were afebrile but still had pain and tightness of the hamstrings. Readings were taken before and after the application of hot packs, ultrasonic, and infra-red radiation. In the 17 patients with neck spasm there was no history of trauma, but radiological examination revealed the presence of osteoarthritis of the spine in 6 of these cases. Before treatment started, the average force necessary to initiate lateral flexion when applied on the painful side was 12 per cent greater than that required when force was applied on the non-painful side, where however the resistance to movement was significantly greater than in the normal subject.

As the treatment of both groups of patients progressed, all three techniques were shown to have produced a significant decrease in the amount of force needed to begin movements, the maximum decline being observed 10 to 15 minutes after a treatment had finished. The effects of hot packs and infra-red radiation on neck

spasm were almost identical and were significantly greater than that of ultrasonic radiation. In the patients with poliomyelitis hot packs were significantly more effective than infra-red or ultrasonic radiation.

It is stated that the difference between the results achieved in normal subjects and in the patients with pain and palpable muscle spasm suggested that static-force balance could provide a 'meaningful measurement of the degree of spasm'. All three forms of therapy produced subjective relief of pain and objective decreases in the amount of force required to move the part [but the results do not substantiate the statement that all sources of heat yield similar effects. For instance, whereas ultrasound produced a rise in temperature at relatively deep levels the main temperature changes produced by infra-red energy and hot packs occurred in the superficial tissues].

A. Garland

Note: The superficial comfort of the hot pack has much to recommend it, despite the proof that ultra-sonic radiation penetrates deeper. Those of us who in former days commonly used hot packs to relieve 'pain or spasm' were in no doubt about their general efficiency. It may still be useful to know that some of the painful phenomena of muscle pain in acute poliomyelitis can be relieved even better by these homely remedies than by infra-red or ultrasonic radiation.—*A. V. Neale.*

Height and Weight of Children with Cerebral Palsy and Acquired Brain Damage

H. M. STERLING. *Archives of Physical Medicine and Rehabilitation*, 1960, **41**, 131–135. 4 figs., 24 refs.

To determine whether the height and weight of children with brain damage differ from those of normal children, the records of 100 children with 'cerebral palsy' were compared with similar records for their 53 siblings. Of the 100 affected children 40 were spastic, 55 were athetoid, and 5 had other conditions, the disability being severe in 50, moderately severe in 30, and mild in 20. Height and weight were charted on the anthropometric charts of the Children's Medical Center, Boston, for ages 6 months to 13 years; for those aged 13 to 18 the NEA-AMA (1949) percentile standards were used. The Wetzel Grid was also used for those aged 5 to 18.

Children with congenital or early-acquired brain damage tended to group in the shorter height and lighter weight areas, while the unaffected siblings were scattered throughout the range, with a tendency towards greater height and weight than expected. Of the affected children, 77 ranked below the 30th percentile of the charts for height and 73 for weight, in contrast to only 4 of 35 healthy siblings in similar age groups for height and 5 for weight. There was a distinct correlation between the degree of disability and the degree of short height and light weight. A group of 6 children with brain damage acquired after one year of age and not later than 7 years of age were also examined; 3 had heights of 90th percentile rank or greater and 4 had weights of 75th percentile rank or greater.

From these findings the author contends that until further detailed metabolic and nutritional studies are carried out on such children the hypothesis of damage to a presumed growth centre or centres is inadequate as an explanation of these findings. *David Morris*

Note: It would need a stretch of the imagination to accept 'growth centre' fault as responsible for subnormal height/weight characters in a high proportion of severely cerebral palsied children. With improving awareness of nutritional needs and methods of feeding such children, their slowed growth is likely to be remedied. All sorts of checks may be present at intervals —periodic simple vomiting, or an unsuspected hiatus hernia, persistent pap feeding and insufficient protein, disturbing superimposed psychological diseases—any or several of these should be kept in mind all through the years of care and management.

—*A. V. Neale*

The Mentally Defective Twin

J. M. BERG and B. H. KIRMAN. *British Medical Journal*, June 25, 1960, **i**, 1911–1917.

The authors have carried out a survey of two series, each of 200 patients consecutively admitted to the Fountain Hospital, London, which admits all varieties of severe mental defectives, and also of a further 121 consecutive admissions to the Ellen Terry Home, Reigate, which admits blind, mentally defective children. Finally, they determined the incidence of twins among a representative group of mental defectives living at home in London. The aim was to ascertain the incidence of members of a twin pair among these groups and to investigate factors related to the observed excess of this incidence over that in the general population.

The incidence of multiple births was 9.9 per cent among the blind mental defectives, 4.0 to 6.0 per cent for other mental defectives in an institution, and 5.3 per cent for those living at home. The estimated expected incidence in the general population is 2.4 per cent at birth and 2.1 per cent in later childhood. The excess incidence of twins among blind over sighted patients was shown to be related to the occurrence of retrolental fibroplasia, and exclusion of patients with this disorder reduced the incidence of multiple births among blind mental defectives to 4.4 per cent.

Examination of birth-weight data showed mentally defective twins to have lower birth weights than either normal twins or single-born mental defectives. The incidence of prematurity in singleton and twin mental defectives was also higher than in single and multiple live births respectively in the general population. Singleton and twin defectives with retrolental fibroplasia had lower birth weights than did sighted defective twins. It was estimated (by Weinberg's method) that 41 per cent of 41 sighted defective twins were monozygotic, and not more than 22 per cent of 9 blind defective twins could have been so.

Four twin-pairs were concordant for mental defect; all were same-sex pairs. Monozygosity was established in two pairs in which the other twin was above average intelligence. In twin pairs containing one normal and one defective member, the latter tended to have the lower birth weight, both in same and in opposite sex pairs. In the 22 pairs of which the defective twin was known to be second-born, 36 per cent of the other twins were stillborn or died in the neonatal period, whereas in the 14 pairs of which the defective twin was known to be first born, 57 per cent of the other twins were stillborn or died in the first few months of life. Thus second-born twins appear to run a greater risk both of early death and of mental defect. An investigation of aetiological factors revealed that in consecutive admissions of 46 twins and 100 singletons, the most striking difference was in the incidence of mongolism (4.3 per cent and 24 per cent respectively). Among other cases, differences in incidence of individual clinical findings were not significant.

R. H. Cawley

Note: Omitting retrolental fibroplasia from this story, it becomes clearer that developmental life (apart from mongolism) in utero is subjected to a multitude of interacting factors for good or ill. The study of the mentally defective twin, with or without blindness or other neurological abnormalities, and where the other twin is (under conditions of monozygosity) normal or even superior in every way, throws up feeling that an enormous amount of study of developing tissues, specially of the nervous system, will have to be made before even the slightest clue of this form of nature's failure is available to us.—*A. V. Neale.*

The Clinical Course, Pathogenesis and Treatment of Epileptic Hemiplegia in Children

V. A. KARLOV. (*In Russian.*) *Pediatrija*, 1960, **38**, 24–29. 26 refs.

From his personal observations of 50 cases of post-convulsive hemiplegia in

children the author, after classifying the disorder under 12 headings, based on the type of onset, the transitory or permanent nature of the hemiplegia, and the subsequent outcome, that is, whether the hemiplegia is progressive or stationary, proposes the following new classification in three main groups.

I. Post-epileptic paralysis. (1) Transitory: (*a*) sudden onset; (*b*) slow onset. (2) Permanent: (*a*) pyramidal; (*b*) subcortical; (*c*) afferent (cases in this last sub-group show lesions of the post-central cortical regions, with sensory changes but no evidence of motor paresis).

II. Paroxysmal epileptic hemiplegia, in which the paralysis is an epileptic equivalent. (1) Type 1, of short duration (less than an hour) and mild. (2) Type 2 lasts some hours or days, during which pyramidal signs are present.

III. Pre-convulsive hemiplegia, in which the paresis appears as an epileptic aura. Cases of Group II are rare, and those of Group III still rarer, only one case of the latter being observed in the 50 patients studied.

Of 26 patients treated by the author, 22 responded, and in some the hemiplegia practically disappeared. Physical treatment consisted of gymnastics and massage, with active movements, while drug treatment included administration of 'Mellictin' (to diminish muscle tone), nicotinic acid, 'Dibasol', glutamic acid, and anticonvulsants. Aetiologically 10 of these patients were suffering from intra-uterine deformities resulting in Weber's disease, congenital hydrocephalus, platybasia, and sclerotic hemiatrophy, in 7 there was a definite history of birth trauma and in 4 others a probable such history, in 6 a history of cerebral infectious disease (meningitis, encephalitis or arachnoiditis), 7 had suffered severe cranial trauma in early childhood, and 2 had been found to have Rh incompatibility at birth. A family history of epilepsy was present in 7 cases.

L. Firman-Edwards

Note: We are grateful for abstracts of Russian papers, especially on such an important subject as hemiplegia in childhood. The breakdown of 50 cases of post-convulsive hemiplegia into 12 varieties on clinical and chronological grounds indicates how variable and almost individually unique each case is. Close study of the clinical features and, whenever possible, correlation with the neuroanatomical pathology should help to improve our very limited knowledge of hemiplegia in childhood. The group called afferent 'with sensory changes but no evidence of motor paralysis' should stimulate more clinical searches for such post-convulsive phenomena.—*A. V. Neale*

H.H.E. Syndrome. Hemiconvulsions, Hemiplegia, Epilepsy. (*In English*)

H. GASTAUT, F. POIRIER, H. PAYAN, G. SALAMON, M. TOGA, and M. VIGOUROUX. *Epilepsia*, June, 1960, **1**, 418–447. 20 figs.

In this paper from the University of Marseilles the syndrome of hemiconvulsions, hemiplegia, and epilepsy (H.H.E.) in infancy is discussed with reference to the clinical and EEG findings in 150 patients and the necropsy findings in 16 of them.

It is pointed out that the H.H.E. syndrome has an essential precursor, the acute illness in infancy which the Marseilles school have termed the hemiconvulsions and hemiplegia (H.H.) syndrome. The typical clinical features of the latter syndrome are sudden onset of convulsions involving one half of the body, continuing from a few hours to several days, and associated with a flaccid hemiplegia which becomes obvious on cessation of the convulsive movements. There is usually a marked pyrexia, and coma often supervenes. There may be papilloedema and the cerebro-spinal fluid is either normal or shows a pleocytosis. The syndrome is most common between the ages of 6 months and 2 years and in the majority of cases appears to be primary with no clear-cut precipitating factor. After recovery from this acute episode, there is a latent period of about one year (with extremes which range from a few weeks to 20 years) before the secondary

epilepsy appears. It is the addition of epilepsy to the sequelae of the H.H. syndrome which constitutes the H.H.E. syndrome. This late epilepsy may consist in unilateral convulsions, as in the original H.H. syndrome, or in convulsions which are generalised from the start. After a long latent interval, symptoms of temporal-lobe epilepsy appear in more than half the cases.

EEG and pneumoencephalographic findings in both syndromes are described. These indicate initial lesions in the affected hemisphere during the acute illness and provide evidence of secondary lesions in the chronic stage (H.H.E.). The authors explain the evolution of these syndromes on the basis of their clinical, EEG, X-ray, and pathological observations as follows. An initial traumatic, vascular, or infective process produces a primary, local hemispheric lesion which is responsible for the initial hemiconvulsions and hemiplegia. This local lesion may resolve or may produce a cicatricial, irritative focus, which gives rise to persistent or even progressive hemiplegia and convulsions. Even if the primary lesion resolves, however, initial gross oedema of the hemisphere (as shown in the pneumoencephalogram) may produce temporal herniation with local ischaemia and cytotoxic oedema, causing secondary lesions that lead to irritative scarring capable of producing late epilepsy, which is often of the temporal-lobe type.

[This comprehensive study contains a list of 141 previous publications on the subject.] *J. B. Stanton*

Note: While the authors 'explain' the evolution of these syndromes it is equally clear that the initial process is often quite mysterious, and the unilaterality is often puzzling. Undoubtedly the train of resultant pathology in a hemisphere is frequently almost inevitably progressive in its damaging effects on normal function and later on cerebral rhythm. One of the most disturbing effects is the involvement, sooner or later, of temporal lobe tissue as a sequel to temporal herniation in the acute and initial disorder. It cannot ever be overemphasised that a convulsive syndrome in infancy, from whatever possible or proven cause, is fraught with serious danger. Urgent therapeutic measures must be available and applied at this stage if some of the H.H.E. disasters are to be avoided or at least minimised—*A. V. Neale.*

Central Haemorrhagic Encephalopathy of Early Infancy

C. B. COURVILLE. *Neurology*, Jan. 1960, **10**, 70–80.

This study is based on 3 cases of acute generalised haemorrhagic necrosis of the cerebral centrum of early infancy. The lesions were so severe as to be incompatible with life for more than a few hours. From clinical information present in each instance, it seems logical to conclude that acute anoxaemia was the primary cause. This conclusion seems reasonable because, in many instances of acute severe cerebral anoxia, numerous petechial haemorrhages are found in the white matter of the brain. The remarkable similarity of the distribution of the acute lesions in these 3 instances to that of the structural changes found in the so-called chronic cystic degeneration of the cerebral centrum in infants raises a question of the possible relation of cause and effect of the two. This possible connection is considered in some detail and the question of anoxaemia or some gross impairment of the foetal circulation accociated with the birth process is reviewed. The most likely answer to this question seems to lie in this possibility.

Author's Summary

Note: These findings indicate again the dangers to special parts of the brain arising from the possible wide and rapid fluctuations in the cerebral circulation at or very soon after birth. The so-called 'border-line zones' in the arterial areas of the brain are particularly vulnerable to ischaemic-anoxic damage. Episodes of very low blood-pressure in the newborn from birth-shock could meet the reasoning put forward by Courville when he notes a possible con-

nection between the instances of acute severe cerebral anoxia with numerous petechial haemorrhages in the white matter of the brain (the cerebral centrum of early infancy) and gross impairment of the circulation associated with the birth process. This is a very important report on a special aspect of cerebral pathology.—*A. V. Neale.*

Influenzal Encephalopathy and Post-influenzal Encephalitis: Histological and Other Observations

J. G. HOULT and T. H. FLEWETT. *British Medical Journal*, 1960, **i**, 1847–1850. 6 figs., 8 refs.

The histological findings in the central nervous system of 4 children of 5–13 years and one adult of 44, who died from encephalitis associated with influenza, are described. The clinical details of these cases have been described by Flewett and Hoult (*Lancet*, 1958, **ii**, 11; *Abstr. Wld Med.*, 1959, **25**, 10). The changes observed in the children's brains were (1) scanty lymphocytic infiltration of the perivascular spaces of the brain and meninges, and (2) microglial nodules replacing isolated Purkinje cells and cells of the spinal grey matter. The findings in the adult were those of acute haemorrhagic leuco-encephalitis. Only in this last case was the diagnosis of influenzal infection not confirmed on virological grounds. *J. B. Cavanagh*

Note: 'Influenzal' disease (the word has an ancient astrological derivation) is occasionally as mysterious and dangerous as the 'epidemic sweating sickness' of the Middle Ages. The nervous system is at times amazingly vulnerable to this virus's action, either direct or through toxic or even allergic influences. These fortunately rare forms of influenzal encephalitis doubtless create chaos in the brain, and even if the patient recovers his brain may remain 'sensitised' and may show strange clinical responses during or soon after subsequent infections. Genetic factors probably play a part in personal idiosyncrasy to influenzal disease of this kind.—*A. V. Neale*

Unrecognised Virus Infections and Foetal Malformations

M. DUMONT. *Presse médicale*, June 4, 1960, **68**, 1087–1089.

The author bases this report on information obtained by personal inquiry from 224 women attending the Obstetric Clinic of the Faculty of Medicine, Lyons. He rejected the use of questionaries as he believes that a woman is never accurate in giving details relating to a pregnancy, and considers that by making personal contact he was better able to confirm the diagnosis of, and to get a definite date for, any contact she had had with a case of rubella (German measles), measles, varicella, or mumps. In each case the degree of probable immunity she possessed to these 4 infections was assessed.

The replies obtained from 224 pregnant women were considered reliable, a number of results being discarded for various reasons. The women were separated into the immune, the non-immune, and doubtful, the latter being the largest group. Exposure to infection occurred 138 times during the first 3 months of pregnancy and 86 times during the ensuing 6 months; the number of malformed babies was 8 in the first group and 2 in the second. These figures did not include babies with malformations which are known to arise from other causes, such as mongolism and anencephaly. The percentage of anomalies arising in mothers at risk during the first 3 months was greatest among those in contact with cases of rubella. It is pointed out that of 8 mothers who were given gamma globulin as a pretection 4 were known to be immune, 4 were classed as doubtful, and that all 8 children were normal (7 mothers being exposed to rubella and one to measles). The author was himself unable to examine all the newborn infants, but information on those not seen by him was available from the social workers, who may not have reported all defects to him. The figures for the numbers of malformed infants therefore erred on the low rather than the high side.

The author stresses that his observations strengthen the view that every effort should be made to protect mothers from virus infections, particularly during the first 3 months of pregnancy.

J. G. Jamieson

Note: Most people now suspect that the entry of a virus into an embryo is anything but a desirable matter. But if we are ever to get exact data on the real incidence there must be much more intense study—clinical and laboratory—of the epidemiology of overt and subclinical infection in the community. After all, we are only in a day-to-day balanced resistance to many viruses: possibly some are resident within us all the time! Obviously, as M. Dumont has again indicated, as good health and hygiene as possible in the first trimester of pregnancy has, in this respect as in many others, immense 'feed-back' to the embryo's welfare and a probable greater maternal resistance to viraemia and hence to the transplacental spread of viruses.—*A. V. Neale.*

Congenital Abnormalities of the Eye with Particular Reference to Prematurity

C. A. BROWN. *Proceedings of the Royal Society of Medicine*, 1960, **53**, 189–192.

Congenital abnormalities of the eye may be connected with prematurity either as an associated developmental abnormality or as an acquired post-natal disease. In discussing this fact, C. A. Brown notes particularly optic atrophy, squint, cataract, chorioretinitis and myopia. However, prematurity does not necessarily do more than increase the likelihood of some ocular defect. Most premature infants' eyes are and remain normal. Visual defects have been recorded in variable incidence, but some have probably no higher incidence in prematures than in a comparable group of mature infants. In cerebral palsy, in prematures or others, there is a significant number of examples of optic atrophy which may add to the overall handicap. Anoxia during or immediately after birth can be a menace to the central nervous system and the ocular structures. Excessive use of oxygen is known to have an aetiological relationship to neonatal retinopathy (retrolental fibroplasia). Special care in the perinatal period is fundamental in the prevention of ocular complications arising out of injury at birth and lack of very skilled attention afterwards.

Brown considers some 'statistics of incidence' in the various ocular conditions mentioned and he finds a slightly or moderately higher incidence of 'squints' in the prematurely-born. Some have found no detectable differences. It may be that at very low birth weights (3 lb. or less) the incidence is greater. There may be a greater incidence of congenital cataract; 29 per cent of cases in a Bristol series occurred in premature infants.

In discussing rubellar cataract, Brown records a prospective enquiry in which ocular defects occurred in 50 per cent of all cases of maternal rubella contracted in the first month of pregnancy and to a diminishing incidence in later weeks to the 12th. The cataract may be unilateral and the infants may be premature. Several series of cases indicate that the most vulnerable age in material rubella for heart defects is 3–8 weeks and for deafness 9–12 weeks. Rubella, mumps varicella or morbilli occurring shortly before pregnancy have no known adverse effects on the eyes or elsewhere. Maternal infections, like toxoplasmosis, may determine ocular disease—e.g., chorioretinitis and hydrocephalus. Microphthalmos may be present. Buphthalmos may be associated with prematurity. High myopia has been found in 2.4 per cent of a series of 375 premature children, compared with 1.3 per cent in mature-born children. Partial retrolental fibroplasia may accompany myopic defects. Eleven myopes, including six high myopes, were found among 43 children with a birth-weight of about 3 lb. Brown records that temporary myopia of 5–20 dioptres is relatively common in small prematures in the early weeks of life, but this usually disappears by 3–6 months. However, some cases fail to resolve. It is not clear why

myopia may persist as a residual ocular anomaly after premature birth.

A. V. Neale

Cold Injury in the Newborn

B. D. Bower, L. F. Jones and M. M. Weeks. *British Medical Journal*, Jan. 30, 1960, **i**, 303–309.

The clinical features of cold injury in the newborn were fully described by Mann in 1955, and since then this condition has been more frequently recognised. A retrospective survey at the Children's Hospital, Birmingham, showed that, between 1946 and 1956, 183 newborn infants were admitted with a temperature of 95° F. (35° C.) or lower. The cause of the low temperature was considered to be primary cold injury in 70 patients (42 male and 28 female), but prematurity (48), cerebral birth injury (23), asphyxia (9), and severe infections (15) were among the other common causes. The symptoms in the infants with primary cold injury in order of frequency were refusal of feeds, swelling of the limbs, coldness to touch, lethargy, vomiting, oliguria, redness of the skin, and jaundice. At the time of admission to hospital the commonest clinical features were oedema, immobility, sclerema, and redness of the face. The most important aetiological factor was a low environmental temperature; the external temperature recorded at Edgbaston Observatory at the time 53 of the 70 infants were admitted was below 35° F. (1.7° C.). However, 7 infants were admitted when the external temperature was 44° F. (6.7° C.) or higher, but the signs and symptoms in these patients were typical of cold injury. Other aetiological factors considered important were insufficient clothing or tight wrapping ('cocooning') of the infant which restricts physical activities.

The prevention of neonatal cold injury demands an awareness of the danger of exposure to cold on the part of all who handle the infants. When the condition is established the aim of treatment should be to disturb the infant as little as possible, to raise the body temperature gradually over a period of several days, and to ensure an adequate oxygen intake; antibiotics should be given prophylactically. Of the 70 infants in this series 18 died; post-mortem examination in 15 revealed pulmonary haemorrhage and infective lesions. Follow-up information concerning 36 of 52 survivors showed that general growth was normal except in one child with acyanotic congenital heart disease; in 2 there was mild mental retardation and in one spastic quadriplegia with gross mental retardation.

R. M. Todd

Note: Cold injury in the newborn is a complex disorder, and despite the (probable) declining incidence there are particular and dangerous possibilities of direct complications—e.g., overwhelming infection, with or without convulsions and pulmonary haemorrhage. In this article data were presented in a series of newborn infants having temperatures of 95° F. (35° C.) or lower and the clinical pictures are discussed. It is noted that there are particular susceptibilities in prematurity, cerebral birth injury and asphyxia towards the 'cold injury' syndrome. Acute infection however may be an *initial* factor in causing the body temperature to fall to a low level. Diagnostic confusion is possible: in any case, whether by cause or effect, acute infection, especially respiratory and staphylococcal, must be anticipated, prevented or treated. The brain may suffer toxic or biochemical injury, but fortunately in the follow-up of the present series a large number of the survivors showed normal growth and development, though in a few there was minor mental retardation and one developed spastic quadriplegia.—*A. V. Neale.*

Postnatal Arrest of Development of the Brain and Its Regression

I. Lesný and F. Dittrich. *Zeitschrift für Kinderpsychiatrie*, Jan.1960, **27**, 1–6.

At the Neurological Clinic, Prague, the authors observed 5 children whose physical and mental development appeared normal up to 3 months to 2 years of age, but then

suddenly ceased, and often actually regressed. This was usually a sequel to head injury or infection. Convulsions and severe neurological lesions, such as spastic quadriplegia, developed subsequently. The first child was 7 months old when he became ill and eventually died aged 21 months after a convulsion. The necropsy showed a hypoplasia of the brain, which corresponded in size to that of a child of 6 months. There were no other lesions. The second child was normal up to 2½ years, but sustained a head injury when one year old. From the age of 2½ he suffered from grand mal epilepsy and progressive mental retardation. At the age of 8 his I.Q. was below 25 and his electroencephalogram showed gross evidence of epilepsy. The third child developed normally up to 2 years when he sustained a head injury. He became grossly retarded, with spastic quadriplegia. He died at 3 years of age. At necropsy premature synostosis of the sagittal sutures and hypoplasia of the brain (which corresponded in size to that of a 4-month-old baby) was found. The fourth child developed normally for 20 months. After a sore throat gradual neurological deterioration followed, with the development of a pontine syndrome. The electroencephalogram was of infantile pattern and air encephalography showed hydrocephalus. At the age of 2½ he was grossly retarded and had spastic quadriplegia and epilepsy. He died at home later. The fifth child developed normally up to 2 years. After an attack of mumps his development became arrested and later regressed. He developed a spastic paraplegia, and air encephalography showed diffuse brain atrophy. By the age of 5 he was blind, unable to sit and had quadriplegia. The electroencephalogram showed a grossly abnormal pattern. The authors believe that these cases are unique in the literature, as similar clinical pictures in the past were usually attributed to prenatal causes.

[This group of cases is not altogether homogenous. No reference is made to 'hypsarrhythmia' or 'infantile spasms,' in which condition normal early development is usually followed by gross intellectual regression not unlike that recorded in this paper.] *John Lorber*

Note: The possible association of postnatal injury and cerebral deterioration must never be over-minimised. On the other hand, fortuitous circumstances may be diagnostically and prognostically misleading in this field. This paper shows the difficulties and at the same time indicates the very severe forms of cerebral deterioration in intellectual and motor functions which may follow commotio cerebri traumatica in infants and young children. At this same age, however, it is well known that the brain may fail for natural reasons and causes—a point made in John Lorber's comments—with possible 'hypsarrhythmia' and intellectual regression. It would therefore be unwise to argue teleologically on an accident cause when it might be a relief for the parents to know that an alleged head injury is not in fact necessarily the cause of the trouble. Moreover, important genetic factors may have to be considered.—*A. V. Neale.*

The Brain and Mental Retardation

L. CROME. *British Medical Journal*, March 26, 1960, **i**, 897–904, 33 refs.

The author reviews the morphological basis of mental retardation as evidenced by the findings at necropsy on 240 mentally deficient patients at the Fountain Hospital and 42 at other hospitals in London. He admits that his material is necessarily heavily biased towards the younger and the more severely handicapped patient, and this is reflected in the high incidence of gross anatomical lesions. A classifiable disease process was present in about one-third (91) of his 282 subjects; 36 of these were mongols and in 21 others mental retardation was due to hypertensive hydrocephalus, while 15 defined syndromes, including tuberous sclerosis, kernicterus, and cretinism, accounted for the rest of this group.

In the remaining 191 subjects no classifiable disease process was present; for the

most part the brain showed a variety of pathological changes, often multiple, and was less than 80 per cent of the normal weight (micrencephaly). In one group there was no detectable structural abnormality other than micrencephaly, yet the mental defect in these patients had often been no less than in others with severe anatomical lesions. In 8 cases the brain was apparently normal both in weight and structure; the author stresses the possible background of biochemical disorder that may exist in such cases, but does not neglect to underline possible environmental factors as well.

The various conditions known to influence the development and maturation of the brain are briefly reviewed and the author emphasises the large gap that exists between the experimental findings in animals and understanding of the causation of the various human disorders of mental development. He points out that experimental and statistical approaches to the aetiology of these disorders are fraught with special difficulties and 'cannot replace the time-honoured careful clinical and pathological case studies'. *J. B. Cavanagh*

Note: Anatomical and biochemical factors in mental retardation are the subjects of much current research. Crome's paper is a good attempt to make use of an exceptional supply of necropsy material. Detection of 'defined syndromes' was possible in much less than half of the 282 cases. This merely goes to show the urgent need for new methods for the detection of perhaps electron microscopic abnormalities in the cerebral tissues and cells. New discoveries may have to await further advances in biochemistry and biophysics. It will not be easy or satisfactory until a research team of experts can get the material as early as possible for suitable analysis.—*A. V. Neale.*

Relationship of Neonatal Apnea to Development at Three Years

C. B. ERNHART, F. K. GRAHAM, and D. THURSTON. *A.M.A. Archives of Neurology*, 1960, **2**, 504–510. 1 fig., 9 refs.

The sequelae of perinatal anoxia was studied in 355 children born in the Maternity Hospital, St. Louis, Missouri, who were examined for the purposes of this investigation when they were 3 years old. Of the 355 children 116 had had perinatal anoxia, 159 were normal at birth (born at full term after an easy delivery with an uneventful prenatal and neonatal course), and 80 who had had such complications as haemolytic disease, skull fracture or intracranial haemorrhage, or who were premature. The criteria on which anoxia was diagnosed were satisfactory. The examination at 3 years was carried out without knowledge of the classification of the infant at birth and include a battery of psychological tests and a neurological examination.

The only significant psychological differences concerned the 'cognitive' or 'intellectual' functions. There was a greater impairment in conceptual ability than in vocabulary skill in the anoxic children, and their I.Q. score was slightly lower than that of the normal controls. In addition there were more abnormal neurological findings in the anoxic group than in the controls. The authors discuss the possibility that of genetic factors being associated with both the perinatal complications and the inferiority found at the age of 3 years.

[This is a carefully planned, controlled study, contributing useful information to knowledge of the sequelae of perinatal asphyxia.] *R. S. Illingworth*

Note: We are assured that the criteria on which anoxia was diagnosed in these children were satisfactory, so that the reader is recommended to go back to the paper by C. B. Ernhart *et al.* and study closely the sequelae at 3 years of age. It is important to note the impairments at the highest levels—conceptual especially. It is in these areas of cerebral inadequacy that we must diligently search for remote disorders from perinatal anoxia. This should be more widely taught and appreciated by all, not least by those responsible for antenatal care and obstetrics and for the immediate care of the newborn.—*A. V. Neale*

Maternal Diabetes. Changes in the Hearing Organ of the Embryo: Additional Observation

G. KELEMEN. *A.M.A. Archives of Otolaryngology*, 1960, **71**, 921–925.

Although it has been known for at least a century that there was some association between diabetes and damage to hearing and vestibular function, the main interest in the past has been centred on the danger of middle-ear suppuration in diabetic patients. In recent times this has been controlled by the use of insulin, sulphonamides, and antibiotics. There remains, however, the problem of the effects of diabetes on the inner ear and particularly those of maternal diabetes on the ear of the foetus. The present author reports the changes found in the inner ear in 2 embryos of women with diabetes whose pregnancies were interrupted on medical grounds by hysterotomy at the fourth and fifth months respectively. (These 2 cases, one of which was previously described by the author (*A.M.A. Arch. Otolaryng.*, 1955, **62**, 357) seem to have been the first embryos of diabetic patients examined histologically.) In each case the outstanding abnormality was a tendency to rupture of the vessels and haemorrhage, with resultant damage to the sensory end-organs of the inner ear, with destruction of cupulae.

[These findings suggest that more attention should be paid to the occurrence of hereditary deafness in the children of diabetic mothers, since the earlier such a condition is noted, the better will be the chance of suitable training for the child.]

F. W. Watkyn-Thomas

Note: The relationship of maternal health to the health and welfare of the embryo-foetus is only beginning to be explored. The suspicion that (plus all the possible troubles of the diabetic mother) the embryo is subject to interference with its inner ear development comes as a moderate shock. This is certainly worth investigating further. We hope it is only a rare phenomenon but the present-day survival of many foetuses of diabetic women render the enquiry increasingly important.—*A. V. Neale*